A Parents' Handbook For Sickle Cell Disease

PART II

Six to Eighteen Years of Age

CALIFORNIA DEPARTMENT OF HEALTH SERVICES

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Sickle Cell Disease

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Preface

If you have a child with sickle cell disease who is between 6 and 18 years of age, this handbook is for you! This handbook is the second part of a series for parents of children with sickle cell disease. Part I was written for parents of children from birth to 6 years of age. This book, Part II, focuses on older children and teens.

By using this handbook and working together with your child's health care provider, you can make sure that your child gets the best care. If you have a special concern, use the Table of Contents to find the chapter that covers this concern. For example, if your 8 year-old child is having a problem with gallstones, you will find that Chapter 4 covers health concerns for school-aged children. Or, if you want to know more about school success for your teen, you will find it covered in Chapter 7. Once you find the right chapter, go through the pages until you find your concern.

We hope that you will share this textbook with others who are close to you. This can help them learn more about sickle cell disease. Relatives, friends, teachers and your family doctor may have questions that this handbook can answer. The more they know, the more they can help you and your child. You don't have to manage all by yourself.

Note: In some of the chapters in this book, your child will be referred to as male. In others, your child will be referred to as female.
ALL of the information applies to both girls and boys unless it is clearly stated.

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George C. Cunningham, MD, MPH, Chief
Genetic Disease Branch
California Department of Health Services

Note to Health Care Providers

This handbook is the second part of a two-part series to assist parents of children with sickle cell disease. It was developed in response to requests from parents for more information about the care of their children.

"A Parents' Handbook for Sickle Cell Disease, Part II" covers the developmental years from school age through adolescence. In addition to ongoing medical needs, the handbook addresses other areas at home, school and in the larger community that have been voiced as concerns by parents. The goal of this second volume is two fold: first, to continue to support parents as partners in the care and the development of their children; and second, to help parents of adolescents to begin to shift some of the responsibility of care to their teens.

The role that parents play in the care of their school-age and adolescent children with sickle cell disease is very important. The intent of the authors is that parents will use this handbook as a resource guide in conjunction with the care provided by a sickle cell center or a primary care physician treating a child with sickle cell disease.

Parents of children with sickle cell disease were involved in the development of this handbook. The formal field test with parents resulted in many comments and suggestions that were incorporated into the final version.

Table of Contents

Key Guidelines for Parents

Most children with sickle cell disease can look forward to a full life. It is important that they be prepared for adulthood and living on their own. School, friends, hobbies and work are all part of helping them grow up.

The outlook for people with sickle cell disease gets better every year. Research on how to treat and cure sickle cell disease is being done. We hope that we will make even more progress during the coming years. With parents, health care providers and researchers working together, children with this disease will be better able to enjoy a full life.

There are a few key guidelines that are important for all parents of children with sickle cell disease. These guidelines are briefly discussed in this section. They also appear in each of the chapters in this book. We hope that you will use these guidelines to help you be the best possible parent for your child.

Guidelines for Parents

- ♦ Prevent Problems
- ♦ Get Problems Treated Early
- ♦ Help Your Child Take Care of Himself
- ♦ Build Self-Esteem
- ♦ Take Care of Yourself and Your Family
- ♦ Get Expert Help When Needed

Key Guidelines for Parents

See Appendices A and B for answers to the questions "What is Sickle Cell Disease?" and "What Causes Sickle Cell Disease?"

Sickle Cell Disease Research

Much of what we have learned about how to prevent and treat problems with sickle cell disease has come from research with people who have the disease. These research projects need your help to meet their goals. You may want your child to take part in one of these projects. When you take your child to his doctor or sickle cell center for exams, ask for more information.

Prevent Problems

The best way to help keep your child healthy is to prevent problems from getting started. There are many different ways to prevent problems. Here are some of the most important ways described in this handbook:

- Keep giving your child penicillin until your doctor says to stop.
- Take your child for routine check-ups even when he is healthy.
- Make sure your child gets all the shots he needs.
- If your child has sickle cell pain, help your child manage it at home.
- Follow your doctor's advice for care at home.

Get Problems Treated Early

Not all problems can be prevented. But most problems can be handled if they are found and treated early. Watch your child for danger signs and call or take your child to see the doctor when needed. Also, take your child to the doctor for routine check-ups.

Help Your Child Take Care Of Himself

Children with sickle cell disease need to learn to do things for themselves. Sometimes, it is hard for parents of a child with a chronic illness to allow their child to grow up. They may feel like they need to protect their child all the time.

Start young. Help your child do things himself. As he gets older, allow him to do more things for himself. By the time he reaches the teen years, he will be able to handle most of his own care. Then, when he is an adult, he should be ready to live on his own.

Build Self-Esteem

There are many ways that you can help your child learn to feel good about himself. These ideas will be covered in more detail in this book:

- Listen to your child. Let him know that you value what he says and does.
- Praise him when he does something well.
- Spend time with him.
- Help him learn to do things that he can do well and enjoy.
- Expect success.

Children who have a chronic illness like sickle cell disease sometimes have poor self-esteem. Help your child see himself as a person, not just a disease. Make sure that you and others treat him as a whole person. Expect him to succeed. Help him overcome any problems that get in the way.

Take Care Of Yourself And Your Family

You need to help your child with sickle cell disease stay strong and healthy. Your other family members also need you to make sure their needs are met. And you need to have a good life, too.

It is often hard to balance each person's needs. It is even harder when your child with sickle cell disease is having problems. Still, this is all part of your challenge. You may need to ask relatives or friends to help out with your family or give you a break.

Get Expert Help When Needed

It is a sign of strength to reach out for help. If you, your child or other family members are having problems, get outside help. There are many sources of help. You can go to relatives, friends or a parent support group. You can also see a social worker or counselor who knows a lot about sickle cell disease.

Routine Health Care and Home Care

Children with sickle cell disease need to take care of their bodies. With special care, many problems can be prevented. You can help your young child do what is needed to take care of himself.

You can take him for well child exams and make sure he gets all the shots he needs. You can work with his doctor to make sure that he gets helpful referrals and tests. You can also make sure that he takes any medicine that he needs.

It is best to help your child do more now so that he will be able to take care of himself when he gets older. When your child is young, you will need to help him do many things. As he gets older, he will be able to do more and more things on his own. The transition to adulthood is difficult for all adolescents with sickle cell disease. Your child should become familiar with the adult program or adult internist before he actually has to make his first appointment. By the time he is an adult, he should be able to take care of all of his health and home care needs.

This chapter covers these health and home care topics:

- ♦ Well Child Exams
- ♦ Shots
- ♦ Common Medical Tests
- ♦ Using Other Doctors (Specialists)
- ♦ Taking Medicine at Home
- ♦ Taking Care of Their Own Health Needs
- ♦ When to Call the Doctor or Nurse

Well Child Exams

How Often Your Child Should See a Doctor

- When your child is 6-8 years old: **Every 3-4 months**
- When your child is 9-18 years old: **Every 4-6 months**
- When your child is over 18: **Every 6-12 months**

Well child exams are essential for your child's health. At these visits, the health care staff checks your child's whole body. They also do hearing and vision exams once a year.

Look at the table to the left to see how often your child should be seen. Older children and adults don't need to see the doctor as often as younger children, unless they are sick or having problems. If your child goes to both a family practice doctor and a sickle cell center, the family practice doctor will do most of his routine check-ups. Your child should go to the sickle cell center two or three times a year for special check-ups, or more often if she has special problems related to sickle cell disease.

Well Child Exam Checklist

At well child exams, the medical staff will check the following:

- ✓ Temperature and blood pressure
- ✓ Heart rate and breathing rate
- ✓ Height and weight
- ✓ Eyes, ears, mouth and throat
- ✓ Head and neck
- ✓ Heart and lungs
- ✓ Abdomen and spleen
- ✓ Skin
- ✓ Penis and scrotum or vaginal area
- ✓ Joints and back

When girls reach puberty, they are also given pelvic and breast exams. When girls become sexually active, they should have a Pap smear once a year.

See Appendix C for a Comprehensive Sickle Cell Disease Care Plan. It describes what should be checked at different ages.

Shots

Timely shots will help keep your child well and prevent diseases. Check with your child's doctor to make sure that he is up-to-date on all his shots.

- **Hepatitis B Vaccine**
Your child should have received a series of 3 vaccines before the age of 18 months. If you're not sure this has been done, talk to your doctor.

- **Flu Shot**
All children and adults with sickle cell disease should have flu shots every year.

- **Diphtheria and Tetanus Booster**
When your child needs these boosters depends on when he had his last shots.

- **Pneumococcal Vaccine Booster**
Whether your child needs this booster depends on when he had his shots and your doctor's advice.

- **MMR (Measles, Mumps and Rubella)**
If your child has not had this shot, he needs to get it.

- **TB (Tuberculosis) Skin Test**
Your child must have this done once a year.

Keep a careful record of the shots your child is given. Bring this record with you whenever you take your child in for health care.

Common Medical Tests

See Appendix D for a description of the Common Medical Tests not described here.

Many of the common medical tests ordered on babies and young children with sickle cell disease are also ordered on older children. Most children will have had these tests by the time they are 6 years old:

- Hemoglobin electrophoresis
- Complete blood count (CBC)
- Reticulocyte (Retic) count
- Kidney and liver function tests
- Urine test
- Blood chemistry tests
- X-Rays

There are a few tests that may be new to older children. These are explained here.

Pulmonary Function Tests (PFT)

These tests check how well your child's lungs are working. Your child blows into a machine that measures how the lungs are doing. Starting at 6 years of age, these tests should be done regularly or when your child has pneumonia or other lung problems.

Pulse Oximetry

This test is done to find out how much oxygen is getting from the lungs to the blood. A plastic band is put on the index finger. A machine then measures how much oxygen is going through the blood. If your child is in the hospital with pneumonia or other lung problems, he will be attached to this machine so the test can be done all the time.

MRI (Magnetic Resonance Imaging)

This may be done to look at the bones of the hips or to look at the brain. It takes pictures with a computer. No X-rays are involved.

Cardiac Echocardiography (Cardiac Echo)

This is a test to determine heart function. It is used to determine whether the heart is working too hard to pump blood through the lungs due to lung damage.

Using Other Doctors (Specialists)

The eye doctor checks the eye for damage from sickle cell disease.

Your child will also be sent to other doctors for special exams. These doctors check your child's eyes and heart.

Ophthalmologist (Eye Doctor)

Starting at age 6, once a year your child should see an ophthalmologist who knows about sickle cell disease. An exam should be done by an eye expert who is also a medical doctor, not an optometrist. The eye doctor dilates the pupil to check for damage from sickle cell disease in the back of the eye. This is not a vision screening.

Cardiologist (Heart Doctor)

If your doctor is concerned about heart problems the heart doctor checks to make sure your child's heart is healthy. Sickle cell disease may cause one side of the heart to become enlarged. The heart may need to work harder than normal and needs to be checked.

See Appendix E for a description of Health Care Providers.

Taking Medicine at Home

Penicillin

Make sure that your child keeps taking penicillin until your doctor says to stop. Whether your child will take penicillin after age 5 depends on your doctor's evaluation. If your child has had a lot of infections or had his spleen taken out, he will most likely need it. Many doctors believe that all children should keep taking it. Research is being done now to find out how long penicillin is helpful.

If your child is on penicillin, he will take 250 mg. twice a day, morning and night. By age 6, most children should be able to take it in pill form. Since the liquid doesn't keep long, it is better for your child to take the pills. Still, a few children aren't able to swallow the pills and have to take the liquid.

As children get older, it is harder to make them take something they don't want to take. If this is a problem with your child, explain why this medicine is so important. If your child still won't take it, tell your doctor. As a last resort, shots can be given.

Other Common Medicines

There are a few medicines that children with sickle cell disease often take at home:

- Tylenol (Acetaminophen) for fever and/or mild pain
- Advil or Nuprin (Ibuprofen) for mild pain
- Tylenol with codeine for severe pain (by prescription only)
- Folate (a vitamin pill taken once a day)

Antibiotics

If your child has an infection, he will probably be given antibiotics. These must be taken until they are finished and there is no medicine left. Even if he feels fine, he needs to take the medicine until it is gone.

Most of the time, antibiotics are taken in pill form. If it is in liquid form, check the bottle to see if it needs to be kept cold.

Antibiotics And Pain Pills Are Not The Same

Antibiotics must be taken for as long as they are prescribed. Even if your child feels better, he should keep taking the pills until all of them are gone.

Pain medicine should only be taken as long as it is needed to relieve pain. As soon as the pain is gone, your child should stop taking it.

NOTE: Don't forget to call your doctor if your child has a fever!

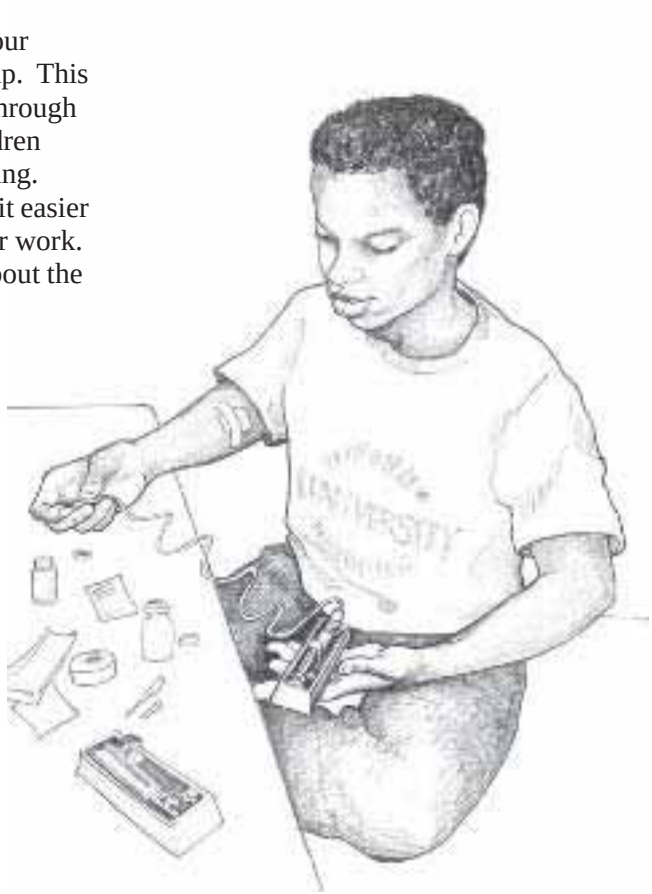
Desferal

Sometimes, children with sickle cell disease need monthly blood transfusions. Chronic transfusions prevent sickling, but they can cause iron poisoning. Desferal prevents and treats iron poisoning.

If your child has chronic transfusions, he must take Desferal. You will need to give your child infusions until he is old enough to do it for himself. At age 10, most children are able to learn to infuse themselves. Make sure your child is taught how to do this.

Desferal is given over an 8 to 10 hour period each day by an infusion pump. This pump slowly lets out the medicine through a needle under the skin. Most children take it at night when they are sleeping. Older children and adults may find it easier to take it during the day at school or work. You can give your child a choice about the timing.

As long as your child gets transfusions, he must take Desferal. Even after the transfusions stop, it can take a while to get rid of all the iron. Your doctor will tell you when your child can stop using Desferal.



At age 10, most children are able to learn to infuse themselves with Desferal.

Taking Care of Their Own Health Needs

Children must play an active role in meeting their health care needs. As they get older, they should depend on you less. Your doctor should ask your child some questions. If not, you should direct the doctor's questions to your child. By the time they are teens, they should be taking care of most of their own health care needs.

Primary School-Aged Children Can Do These Things:

- Learn the danger signs and tell you when they have one.
- Take their medicine (you still need to track how much they take).
- Speak up at doctor visits by asking questions and giving answers about themselves.
- Drink fluids when they are thirsty and at other times when they need more fluids.
- Take breaks and rest when they feel tired.
- Learn to manage mild pain by taking Tylenol, drinking extra fluids, using "home remedies," staying busy or relaxing.

Your teen can talk to clinic staff.

Teens Can Do These Things

- All of the things that younger children can do.
- Call the doctor if they have a danger sign and tell you about their concerns.
- Take their own medicine and tell you how much they are taking.
- Take their own temperature.
- See the doctor while you wait in the waiting room.
- Go to doctor visits without you.
- Manage mild pain by themselves.
- Go to a teen support group.
- Talk to a genetic counselor.
- Keep you informed about what they are doing and how they are feeling.

Learning About The Disease

Talk to younger children about sickle cell disease so that they can tell others about it. Older children can write a report for school, read books and ask their health care providers for more information.

Taking Medicine

Younger children should know what medicine they take and be able to remind someone when they need it. Teens are able to take most medicine on their own. At any age, you need to keep track of the medicine your child takes.

Nutrition

Your child needs to make sure that he eats a well-balanced diet. Children with sickle cell disease need to eat good foods full of protein, vitamins and minerals every day. These foods will help them build new red blood cells. They also need to take 1 mg. of folic acid daily.

Drinking Fluids And Taking Breaks

Children with sickle cell disease need more fluids, bathroom breaks and rest than other children. When your child is away from home, it may be hard for him to take care of these needs. You or your child should tell teachers and other adults about his special needs so it will be easier for him.

Allow your child to take care of these needs himself as he gets older. Check to see how he is doing, but don't follow him around. Help him think of ways to make sure he rests when he is tired. Help him to understand that if he doesn't take care of himself, he may have more problems.

Extra Fluids Are Needed:

- With a fever
- With pain
- When it is hot outside
- With a high level of activity
- When traveling

When your child needs extra fluids, the amount needed every 24 hours is based on your child's weight.

At 40 lbs: Drink 7 cups
At 50 lbs: Drink 8 cups
At 60 lbs: Drink 9 cups
Over 60 lbs: Drink 10 or more cups

A cup is 8 ounces of fluid. The fluids should be clear fluids, like water, bouillon, or fruit juices (not fruit flavored soda).

Taking Trips

See Appendix F for a sample Travel Letter.

Travel is fine for children with sickle cell disease. There are just a few rules for your child to follow which will help him stay healthy:

- Drink plenty of fluids while traveling.
- Fly only in pressurized planes.
- At elevations over 5,000 feet, drink extra fluids and rest often.
- Carry a "Travel Letter" from the doctor.

Watching For Danger Signs

The sooner you find out about health problems, the better. Sometimes, early treatment can save your child's life. Other times, it keeps a problem from causing damage. The longer your child waits to be treated, the worse a problem can get.

Both you and your child can watch for early signs of problems. These danger signs are listed on the next page. Go over them with your child so he will also know what signs are important to share.

Your child needs to drink plenty of fluids.

When to Call the Doctor or Nurse

Call to Have Your Child Seen Right Away

If Your Child Has One of These Danger Signs:

Fever:	101^0 F or higher
Head/Neck:	Severe headache or dizziness Stiff neck
Chest:	Pain or trouble breathing
Stomach:	Severe pain or swelling
Color:	Loss of normal skin color, very pale or gray
Penis:	Painful erection
Behavior:	Seizures Weakness or paralysis (can't move arm or leg) Can't wake up

If you can't reach the doctor, take your child to the Emergency Room.

Call For Advice

If Your Child Has One of These Problems:

Stomach:	Vomits more than once Has diarrhea more than once
Fever:	100^0 F which lasts more than 24 hours
Color:	Jaundiced (eyes or skin look yellow)
Arms, Legs, and Back:	Pain, with no other symptoms
Chest:	Coughs, without fever or chest pain
Nose:	Runny or stuffed nose
Behavior:	Acts strangely Refuses to take medicine Refuses to eat or drink Less active than usual

See Appendix G for a copy of this list to post in your home.

How to Deal with Pain

Pain is common in children and adults with sickle cell disease. Most pain can be handled at home. But if the pain doesn't go away or gets too strong, your child may need to go the clinic or hospital.

As soon as the pain starts, your child should drink lots of fluids and take Tylenol. She can also try other home remedies for pain.

If the pain gets worse or doesn't get better, call your doctor. Your doctor may ask you to find out if your child has a fever. The doctor may also tell you to give your child more to drink, to try heat packs or to use other medicines.

If the pain is too severe, your child may need to go to your doctor's office or the Emergency Room. Call your doctor's office before you take your child to either place so your child will be seen as soon as you get there.

This chapter will cover:

- ♦ Easing Pain at Home
- ♦ Hospital Care for Pain
- ♦ Pain Medication
- ♦ Teaching Your Child to Relax
- ♦ Describing the Pain

Easing Pain at Home

Home Remedies

There are many things your child can do at home to ease sickle cell pain. She can try any of these:

- Drink lots of fluids.
- Rest or play quietly.
- Take a warm bath.
- Put a heating pad or warm, moist towel on sore places.
- Massage the place that hurts.
- Take Tylenol (Acetaminophen) or Advil or Nuprin (Ibuprofen).

Keep Your Child Busy

Keeping your child busy is an important way to deal with pain. She can talk to people, watch TV, play games or listen to music. Let her pick what she wants to do to keep her mind off the pain.

Try not to leave your child alone when she is hurting. Make sure that someone is with her to help keep her involved in other things.

Help Your Child Learn To Relax

Your child may hurt less if she is able to relax. You can help your child learn to relax when she is in pain. With practice, she may be able to stay calm and relaxed. Turn the page to find out how to teach this to your child.

Clinic & Hospital Care for Pain

Your child can control her own pain medicine with a PCA pump.

At the clinic or hospital, your child may get fluids through an IV (in a vein). She may also get stronger pain medicine through an IV or a shot. This helps most children feel better quickly so they can go home. At home, the pain may last a few more days, but it should keep getting better. Again, use fluids, Tylenol, relaxation and other home remedies to help ease the pain.

If the pain does not get better, your child may need to be hospitalized. She will be given more fluids by IV and more pain medicine. Physical therapy may also be used to help with pain. This can include whirlpools, massage, exercise and heat packs. After 3 to 5 days in the hospital, your child should feel well enough to go home.

PCA (Patient Controlled Analgesia)

At many hospitals, your child will get a PCA pump so that she can control her own pain medicine. With a PCA pump, your child can decide when she needs more medicine. She presses a button to pump the medicine into her veins. The pump is set up so that she gets the right amount but not too much medicine. This gives her better control of the pain in a safe way.

Pain Medications

TENS (Transcutaneous Electrical Nerve Stimulation)

A TENS unit may help block the pain. It is a small device prescribed by the physical therapy.

Your child can use it at home when she starts to feel sickle cell pain.

A TENS unit may help to relieve pain.

If your child goes into the clinic, day hospital or emergency room for the treatment of severe pain, she will most likely get strong pain medication. The most common ones are acetaminophen with codeine, morphine, keterolac, and Dilaudid (hydromorphone). Demerol (meperidine) is no longer recommended to treat sickle cell pain.

If your child needs oral pain medication, you will get a prescription from the doctor for a small amount. These drugs should not be stockpiled at home. Also, these drugs (like all medicines) should not be shared with other family members.

Getting the Right Dose

The right dose of pain medication can be very helpful with severe pain. If your child's pain is not better, she may not be getting enough. On the other hand, too large of a dose can cause problems, like sleepiness or pneumonia. If you are concerned about how much pain medicine your child is getting, talk to the doctor.

Side Effects

Pain medications can have some side effects. Often, they cause constipation. If this happens, give your child something to help soften her stools, such as prune juice. Feed her a diet high in fiber, with lots of fruit, whole grains and beans and give her a lot of fluids. If the constipation goes on, call your doctor for advice. Other side effects include itching and mood changes. Pain medication can cause serious problems, but these are rare.

Pain Medication Use Does Not Cause Addiction

Pain medications for short-term sickle cell pain are not addicting. Many parents worry about whether their children will get addicted. Short-term use of these drugs to relieve pain will not lead to addiction.

Addiction Stems From Other Problems

Children with sickle cell disease are not more likely to abuse drugs than other children.

In most cases of addiction, other problems besides sickle cell pain are involved. A teen or adult who is having problems with home, school, or friends may abuse pain medication. When this happens, it is likely that other drugs would be used if the pain medication wasn't around.

Prevention Begins at Home

Speak to your social worker or nurse if you are concerned about drug abuse. Take action early if you think your child is having problems at home or in school. Get involved with drug education programs at school and in your community.

Chronic Pain Syndrome

Pain medications are only for acute pain. They are not meant to be used for chronic, long-term pain. People with chronic pain need to learn other ways to control their pain besides taking pills. Some special pain problems require different medications that your doctor will talk with you about.

Fewer than 1 in 20 people with sickle cell disease have chronic pain that may result in the frequent use of pain medication. If your child is one of these, she should see a team of pain experts for help. This team may include a psychologist, social worker, physical therapist, neurologist and pain medicine expert. The team will draw up a treatment plan for home and the hospital. It is important to follow all parts of the plan.

Teaching Your Child to Relax

Relaxation is a skill that can be learned. Like any other skill, it takes practice to learn to relax. Pick times that are quiet and free of distractions to help your child practice. Bedtime is often a good, quiet time. It is also a time when you may have a few extra minutes to help lead your child through these exercises.

Deep Breathing

Deep breathing is one of the simplest and best ways to relax. Try to find fun ways to help your child to breathe deeply and evenly. Have your child take a few deep breaths and let them out to the slow count of 1…2…3… Or have your child pretend to be a bicycle tire that the air is going out of slowly.

Try one of these ways to keep the deep breathing going for a while. Tell her to pretend that she can breathe under water if she breathes in this special way. Or she can pretend to be an astronaut in a space suit who has to breathe deeply and evenly.

You can help lead your child through relaxation exercises.

Exercises for Younger Children

Exercise #1: The Rag Doll

"Pretend that you are a robot (or wooden doll), all stiff and straight. Your arms and legs don't bend at all. They just stay straight."

"Now you are a rag doll, all floppy, with no bones." (Lift your child's arm up, shake it a little to make sure it's really loose.) "All loose and floppy."

"When you need to relax, pretend to be the wooden doll, then the rag doll."

Exercise #2: Spaghetti

"Pretend that you are spaghetti in a package that has not yet been opened. You are all stiff and straight." (Have your child hold this for a few moments.)

"Now you're cooked spaghetti, all over the plate. Are you covered with sauce or meatballs?"

"When you need to relax, pretend to be spaghetti in the package, then the cooked spaghetti."

Exercise #3: My Special Place

Begin with deep breathing practice. "Each time you breathe out, you get more and more comfortable. Now you're loose and comfortable. No tightness anywhere, breathing deeply and evenly. All your muscles are smooth and warm. Just an easy feeling. As I slowly count backwards, you are going to feel even more relaxed: 5…4…3…2…1…"

"Now imagine that you're in your favorite place (maybe floating underwater or in a made-up land), feeling free and easy. What do you see? Hear? Taste? Feel? Smell?" (It's best if her eyes are closed, but they don't have to be.)

Exercises for Older Children

Begin with deep breathing practice. "Start at the top of your head. As you breathe out, your scalp feels loose and comfortable. Now go down through your face and feel all the little muscles smooth out. Now feel any tightness flow out of your shoulders, down through your arms and hands, out of your body."

"Now feel any tightness flow out of your chest as you breathe out. Your stomach muscles feel loose. You feel any tightness, any tension, flowing out from your hips down through your legs, out the bottoms of your feet. You feel loose and comfortable, more and more relaxed each time you breathe out."

"Now I'm going to count backwards from 5 to 1, and you will feel even more relaxed. 5…breathing deeply and evenly 4...more and more relaxed 3...further and further 2...loose and comfortable 1." (If your child wants, she can imagine her favorite place at the end of the count of 1.)

Teaching Tips

- **Keep your voice low and soothing.** Speak slowly, but not in a monotone.

- **Keep it natural**. These are just outlines of what to say. Say what feels good to you. Soon, you will find your own words for the same things.

- **Work with a psychologist.** It can be helpful to learn these exercises from a psychologist. He or she will pick the ones that will work best for your child and teach them to both of you.

- **Make tapes of the exercises.** A psychologist can also make a tape of these exercises for your child to use as a guide. She can listen to it when she is in pain and can't relax on her own. You or your child can also make your own tape. Add music or a story if it helps.

- **Keep it positive.** Your child does not have to try to relax all of the time. Make it enjoyable so that she'll be willing to try.

Self-Hypnosis

A psychologist can teach your child self-hypnosis. This is a more detailed relaxation technique that your child can use on her own.

What To Expect

It is important for your child to practice these techniques when she doesn't have pain. This practice will help give her the best results.

Relaxation techniques can help your child manage pain. If she feels pain coming on, she should drink extra fluids, take Tylenol and relax in whatever way works best for her. Relaxation exercises and other home remedies alone will often take care of the pain. Other times, medicine may also need to be used.

Your child can listen to a relaxation tape.

Describing the Pain

Figure 1. Pain Scale

You and your child must know how to describe her pain so she can get the right treatment. There are many ways to describe pain. Stick to the one that works best for your child. Make sure her doctor uses the same one too.

• **How severe is the pain?**
First, she needs to be able to say how severe it feels. She can give the pain a number from 1 to 10, with 1 as the mildest pain and 10 as the worst pain. She can also pick a face, from a big smile for no pain to a big frown for the worst pain. The pain rating scale below shows these different ways to rate pain.

• **Where is the pain?**
Her doctor will also want to know where she feels the pain. Sometimes it is easiest to describe where it is. Other times, it is helpful to mark the places on an outline of the body.

• **How does the pain feel?**
Pain may feel sharp or dull, hot or throbbing. If your child can't tell you how it feels, sometimes she can show you with color. Ask her to color how the pain feels and you can show it to her doctor.

Please circle the face that best describes the way you feel right now.

Figure 2. Pain Scale

Living with Pain

Pain Itself is not a Cause for Panic

While sickle cell pain hurts, it rarely causes serious problems. Most of the time, it can be managed at home. This pain is usually not a sign of something worse. It is part of the disease and needs treatment.

You and your child need to know when you should call your doctor with pain. Call your doctor if your child has any of these:
- Chest pain
- Severe headaches
- Severe pain in the belly
- No relief after trying "home remedies"

Not all pain may be caused by sickle cell disease. If you have questions about the cause of your child's pain, ask your doctor.

Get Help With Pain Problems

If your child has many problems with pain, counseling may be helpful. Counseling can help your child learn to manage the pain. A counselor can also help other family members cope. One person in pain can put a strain on the whole family.

Don't let pain take over your child's life. Help your child find ways to ease her pain and to live with it. She will learn more about when she needs to manage her pain and when she can ignore it. Also, ask your doctor about hydroxyurea.

Primary School Years

Children between the ages of 6 and 12 can have most of the problems that affect younger children. The most common problems are:

- ♦ Infections
- ♦ Acute Chest Syndrome
- ♦ Anemia (Low Blood)
- ♦ Gallstones
- ♦ Spleen Problems
- ♦ Priapism
- ♦ Delayed Growth
- ♦ Problems With Kidneys and Urine
- ♦ Strokes and Other Brain Problems
- ♦ Meningitis

In this chapter, we will describe these problems and how they affect primary school-aged children.

Different centers and doctors may use other approaches to treat these problems. Follow the doctor's advice. If you are concerned about what you are told, get a second opinion from another doctor.

Infections

If your child has a fever of 101^{0}F or more, call your doctor's office.

As your child gets older, his body will get stronger. He will be able to fight infections better. He won't need to go to the hospital for infections as often. He can be treated at home and get well.

Still, some infections can cause problems. You still need to watch out for fevers. **If your child has a fever of 101^0 F or more, call your doctor's office.**

See Appendix H for a Temperature Conversion Chart. It shows you how to convert Celsius to Fahrenheit.

Meningitis

This is a severe infection of the membrane that surrounds the brain and spinal cord. It can be caused by a bacteria or virus.

These are the most common signs to watch for in older children:

- Fever
- Severe headache
- Throwing up
- Stiff neck (not able to touch chin to chest)

If the doctor thinks that your child might have meningitis, a spinal tap will be done to test the spinal fluid. If there is an infection, he will be put in the hospital right away and given antibiotics by IV.

Sometimes, after children get over this infection, they may have some hearing problems. Hearing tests should be done to check for these problems.

Acute Chest Syndrome

Acute chest syndrome is the term used to describe pneumonia in sickle cell disease patients. It can have many causes including infection or sickle cells lodged in the lungs. It frequently develops during a pain episode and can be accompanied by low oxygen in the blood.

Early treatment will keep it from getting worse. Watch for these warning signs:

- Fever
- Coughing
- Rapid breathing
- Shortness of breath
- Difficulty breathing or "grunting"
- Severe chest pain

If you see any of these signs, take your child to the doctor right away. Call first and describe the signs your child is showing.

Treatment for acute chest syndrome is given in the hospital. Your child will be given antibiotics and may need oxygen. A blood transfusion may also be required. Most of the time, this treatment makes people feel better.

The doctor may order a chest X-ray every day in the hospital as well as within a few weeks after discharge. The doctor may also repeat lung tests and an echocardiogram in a month or two to see how well the lungs have healed.

Acute chest syndrome can be brought on when a child does not take deep enough breaths. This can happen when too much medication is given for pain or surgery. While taking drugs for pain, your child should expand his lungs by blowing into a balloon or an "incentive spirometer." This is a mouthpiece that shows how hard he is blowing.

Gallstones

About a third of children with sickle cell disease have gallstones by the age of seven. Many others develop them later.

Gallstones in sickle cell disease are formed from the products of broken-down red blood cells. This collects in the gall bladder and forms thick sludge or stones. Gallstones can become stuck in the bile duct where they can cause serious problems by stopping the flow of bile. This requires emergency procedure to remove the stones. After recovering from this procedure, the gall bladder is usually removed.

Signs and Treatment

Often, there is a warning before gallstones get stuck in the duct. When the stones pass through the duct, they may cause pain in the right side of the abdomen. If the gall bladder is taken out after this warning, serious problems can be avoided. When gallstones are stuck in the duct, a person's skin and the white of the eyes may become very yellow.

Your doctor may suggest surgery to remove the gall bladder before an emergency happens. Taking out the gall bladder is the most common surgery in people with sickle cell disease. People can get along well without a gall bladder. However, they may have trouble eating a lot of fatty foods at one time.

Anemia (Low Blood)

People who have sickle cell disease have fewer red blood cells than normal. They often become tired more quickly than people with normal blood counts. In general, people with SS disease have the most severe anemia.

There are times when your child's blood count may fall much lower than usual. This can happen with a fever or an infection. Either the body stops making new cells or the cells are destroyed quicker than usual. When this happens, the destroyed red blood cells in the body fluids can make the eyes look more yellow and the urine look darker.

Signs

Bring your child to the doctor to have his blood count checked if you notice any of these signs:

- More tired than usual
- Pale color
- Loss of appetite
- Yellow eyes or skin
- Dark urine
- Enlarged spleen

Treatment

If your child's blood count falls very low, a blood transfusion may be needed. An extremely low blood count can result in heart failure and death if not treated in time. Since the blood count often falls when there are other medical problems, your doctor will usually check it daily if your child is in the hospital.

Bring your child to the doctor for a blood count.

Spleen Problems

The spleen is in the left upper corner of the abdomen, just under the edge of the rib cage. The spleen's job is to filter out damaged red blood cells from the blood and to help fight infections.

Enlargement and Scarring

The type of sickle cell disease a person has affects the spleen differently. With sickle cell anemia (SS) and sickle beta^0 thalassemia, the spleen often becomes small by age 6, after being enlarged for a few years. Children with other types of sickle cell disease (see Appendix A) can have an enlarged spleen for many years. Sometimes the spleen must be removed due to chronic anemia

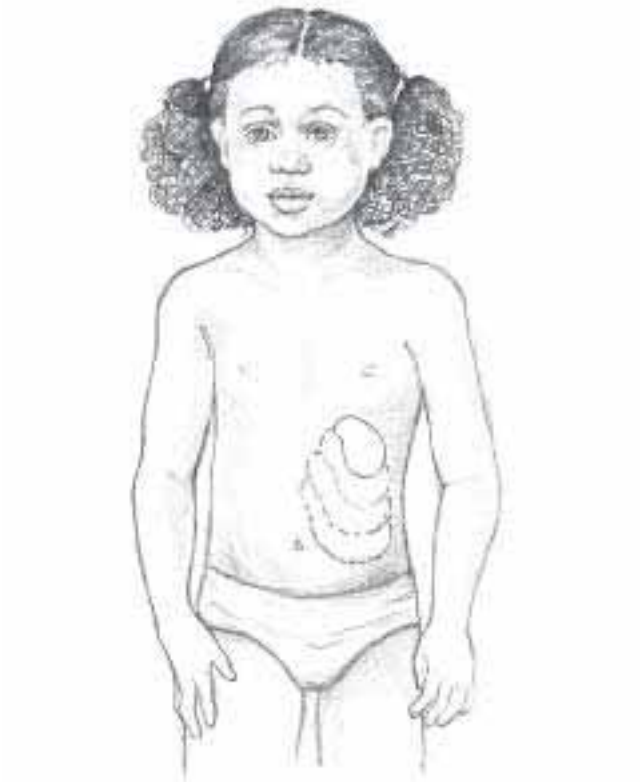
A mildly enlarged spleen doesn't usually cause problems. Still, the scarring keeps the spleen from working well. This is one of the reasons that children with sickle cell disease get so many infections.

Splenic Sequestration

In some children, the spleen may become larger very quickly and begin to trap lots of blood cells. This "bleeding into the spleen" can cause the blood count to drop quickly. When this happens, it is called a "splenic sequestration crisis." It can lead to heart failure and death if not treated promptly with a blood transfusion.

Splenic sequestration can happen when your child has a fever or a cold. Other signs to watch for are abdominal pain or swelling, pale color and fatigue.

If your child has one splenic sequestration, he will be more likely to have another one. His spleen may be taken out so that it can't happen again. He doesn't need his spleen to live. To help prevent infections, he should keep taking penicillin twice a day until his doctor tells him to stop.



Your child's spleen may be larger than a normal spleen.

Priapism

Priapism happens when sickle cells block the blood vessels in the penis. This makes the penis hard and sore. This is different than a normal erection. It can happen at any age.

Most often, boys or young men awake in the middle of the night or the morning with a painful erection that does not go away with time. If your child has a painful erection that does not go away within 30 minutes, call your doctor. He may need treatment right away with a blood transfusion, IV fluids, and pain medication. Sometimes a procedure is required to restore normal blood flow to the penis.

Priapism can occur frequently, but resolve without treatment. This can be as serious as a single severe episode and needs to be reported to the doctor. Your son needs to tell you if he has priapism. Boys often feel shy about telling their parents about this problem. Talk to your son about this before it happens. Tell him that he must tell you if his penis gets hard and sore, even if the pain stops. Even if the priapism goes away, tell the doctor about it. There are medications that can be taken for this problem.

Delayed Growth

Some children with sickle cell disease are small for their age. As they become adults, most children with sickle cell disease reach full size. If your child is smaller than his friends, tell him that he will most likely catch up. It will just take him a few years longer to grow.

Some children with sickle cell disease also reach puberty later than other children. Talk about this with your child early, before he brings it up. Help him find ways to feel good about himself now. Reassure him that he will mature in time.

Some children with sickle cell disease are small for their age.

Problems with Kidneys and Urine

Dehydration

Sickle cell disease can damage the kidneys. Children with sickle cell disease pass urine more often than other children. Therefore, they need to drink more fluids to avoid dehydration. When a child becomes sick and drinks less than usual or loses more fluid by vomiting, diarrhea or fever, he can get dehydrated.

Your child should drink plenty of fluids.

These are some signs of dehydration:

- Urinating much less than usual
- Darker urine
- Difficulty urinating
- Burning when urinating
- Dry, sticky mouth and lips
- Sunken eyes

Sometimes, the skin may feel different. If you pinch it, it doesn't return to normal right away.

If you notice any of these signs, call your doctor and give your child plenty of extra fluids to drink.

Bedwetting

The need to urinate can often lead to bedwetting. This is common in many older children with sickle cell disease.

There are several ways you can help your child stop wetting his bed. It may help to wake your child to urinate twice during the night. This could be just before you go to bed and one other time. You could try setting an alarm clock to go off in the middle of the night so your child can get himself up to go to the bathroom. Your doctor or nurse may have other helpful ideas. Children who are sound sleepers will have more trouble getting up at night to urinate. They may have bedwetting for a longer time. Do not criticize your child for bedwetting.

Kidney and Bladder Infections

With sickle cell disease, bladder infections are fairly common. If they are not treated promptly, they can move from the bladder up to the kidneys and cause kidney damage.

Call your doctor or nurse if you notice any of these signs:

- An increase in the number of times your child goes to the bathroom
- Bedwetting after your child has stopped wetting the bed
- Being unable to hold the urine
- Foul smelling or cloudy urine
- Fever
- Burning and pain when urinating
- Abdominal or back pain

If your child has a bladder infection, he will need to have his urine tested again from time to time. If the infection returns often, he will need to take pills every day so that the infections won't keep coming back. He may need tests to be sure his kidneys have not been damaged.

Blood in the Urine

Another problem caused by sickle cell disease is blood in the urine due to bleeding from the kidney. When this happens, the urine usually looks bright red or brownish. In more severe cases, there may be back pain and small pink specks or lumps in the urine. The blood usually leaves the urine within hours. In some cases, the bleeding can go on for days and become a serious problem.

Always call your doctor right away if you notice blood in your child's urine. Tests can then be done to find out what is causing the bleeding. In most kinds of kidney bleeding, it is very important to get plenty of fluids, sometimes through an IV in the hospital, and to rest in bed. Tests to see if there is blockage from clots may need to be done.

Strokes and other Brain Problems

One of the worst problems caused by sickle cell disease is strokes. Early treatment can help prevent serious damage.

A stroke occurs when part of the brain doesn't get as much blood as it needs. This is caused by sickle cells blocking a blood vessel in the brain.

Watch for these signs of a stroke:

- Sudden weakness or tingling of an arm, leg or the whole body
- A difference in the way one side of the face or one eye moves compared to the other side
- Seizures (shaking that can't be stopped)
- Speech trouble
- Sudden, strong headache
- Fainting

If you see any of these signs, call your doctor and bring your child to the hospital right away. The sooner he gets help, the better.

Minor Strokes and Other Brain Problems

Minor strokes and other brain problems can happen without your child having any signs or symptoms. The only way to know that a small stroke has happened is through some special tests.

Minor strokes often cause learning problems. These learning problems can affect how well your child does at school.

Tests to Learn About The Brain

There are some tests that check whether your child is having problems with his brain due to cells sickling. The results of these tests will tell you if your child needs to take special steps to prevent strokes.

- **MRI (Magnetic Resonance Imaging)**

An MRI is done to look at the brain. Pictures of the inside of the brain are taken by a computer. It shows where an injury is in the brain. It usually takes about an hour and doesn't hurt. Young children may need to be given medication to relax while it is being done since they have to lie still for an hour. No X-rays or needles are used.

- **CAT Scan**

When an MRI is not available, a CAT Scan can be used instead. It uses X-rays to make a computerized picture of the brain and check for bleeding.

- **Transcranial Doppler (TCD)**

Transcranial Doppler is a machine that uses ultrasound to detect areas of increased blood flow in the blood vessels of the brain. When blood vessels are narrowed due to sickle cell damage, the blood makes a louder noise as it travels through the narrow area. If there is faster flow, that means that the blood vessel may be narrower, and there is a greater risk of possibly having a stroke in the future.

Treatment for Strokes

If your child has a stroke, he will need monthly blood transfusions. These transfusions help prevent more strokes. They are usually given for at least five years.

To help your child recover from a stroke, take him to a center that has all of these services:

- Rehabilitation program, including physical, occupational and speech therapy
- Neuro-psychological testing and care
- Desferal instruction

Treatment for minor strokes is often the same as treatment for other strokes. Transfusions are used to help prevent more strokes. If there are learning problems, steps can he taken to help him manage them.

Strokes and other brain problems can cause learning problems in some children with sickle cell disease. To find these learning problems early, all children should be screened at routine exams, starting at age 6.

Transfusions

There are two types of transfusions your child may need. Simple transfusions are the most common. For a simple transfusion, a person is given a set amount of blood through an IV. This is done to treat anemia. For an exchange transfusion, a person is given a set amount of blood while the same amount of blood is taken out of the body. This is done to reduce the amount of sickle hemoglobin.

Each time your child needs to be transfused, his blood will be typed and crossmatched (see glossary) before the transfusion. Complications are rare, but can happen. Complications include rashes, itching, and rarely infections.

Your child may need monthly blood transfusions.

Learning Problems

Any child, with or without sickle cell disease, can have learning problems. Learning problems can affect how well children do at school and in their social lives. Most children with learning problems have normal intelligence. They are just as smart as other children.

There are many different kinds of learning problems. Some children have a hard time taking in information. Others have problems understanding or remembering things. Some children have a problem with reading, writing or other skills.

If your child is having trouble at school or socially, work with the school and tell your doctor. If there is a concern, the doctor will refer your child for neuro-psychological (neuro-psych) testing. This testing is done by a psychologist who has special training in learning problems.

Neuro-psychological testing will help find out how your child learns.

Neuro-Psychological Testing

To do this testing, the psychologist will talk to your child and ask him questions. Sometimes your child will be asked to read questions and write down the answers. Nothing is done to your child's body.

The goal of this testing is to find out how your child learns. The results will help show you and your child's teachers what kind of help he needs.

Help for Learning Problems

A child with learning problems can learn when he is taught in a way that works for him. Often, small changes in how a child is taught can help him do well in school. Also, teachers may be able to show him ways to use his strengths to learn better.

Make sure that you understand your child's problems and how he learns best. Use that information to guide you when you are teaching your child something. Also, stay involved with your child's teachers and school to make sure that they give him whatever help he needs to do well. Many children will need a special program sometimes called an Individual Education Plan (IEP).

What can you do if your child does not qualify for special education, but you know he needs some adjustments to the regular school environment because of his sickle cell disease? The answer is to request a "504-Plan hearing" at your child's school. (For more information about 504-Plans please see Appendix O).

Guiding your Younger Child

While you need to care for your child's health, you also need to focus on more than her health care needs. When your child starts school, she begins a new stage in her life. She will spend a lot of time away from you, in the care of other adults. She will also spend more time with other children her own age. Both her social life and her school life may raise new questions or problems.

Transitions involve change: adding new expectations, responsibilities, or resources, and letting go of others. Transitions, large and small are a predictable part of child and family life. There are transitions that are normal for development, from infancy, to childhood, then adolescence and adulthood. These developmental transitions occur for children with and without chronic illness. Other transitions that all children face include changing from preschool to grade school, then middle school, high school and college or work. Still other transitions include moving into new programs, working with new agencies or providers.

You are the most important person in your child's life and are the key to her success and self confidence. This is a time to help your child learn to do more things for herself. It is also a time to help build your child's sense of herself and to give her a good start with school. These are the same challenges faced by parents of all children, and the same guidelines apply. The goal of your child's health care team is to support you in making your child's life as normal as possible.

This chapter will give you information about how to help your child with these issues:

- ♦ Doing Things For Herself
- ♦ Self-Esteem
- ♦ School Success

It will also help you look at the needs of your whole family and how you can take care of yourself.

Doing Things for Herself

Sickle Cell Summer Camp

Summer camp is always filled with outdoor fun, new friends, singing, swimming, story-times and games. At sickle cell summer camps, counselors know how to help children with the disease do as much as they can, without hurting themselves. The staff is aware of the special needs of children with sickle cell disease and can handle any problems that may arise.

Sickle cell summer camp provides a safe and fun setting for your child to gain confidence and self-esteem. Spending time away from parents with other children her own age will help your child be more independent.

Ask your doctor or local sickle cell group about where you can get information about the nearest camp.

Your child may be old enough to make his own lunch.

Your child can learn to take care of herself, even though she has sickle cell disease. She needs to do things on her own. You still have to see that your child gets what she needs. But your job as parent is changing. You need to help your child learn to do things for herself.

By age 6, most children want to start doing things themselves. Your child may want to:

- Help make her own breakfast or lunch.
- Dress herself and get ready for school.
- Take a class or learn a skill, like playing the piano.

Besides the things she wants to do, she can also help out with the house. She can:

- Help with household chores.
- Clean up her room.
- Clear and set the table.
- Do the dishes.

Let her do things herself unless you're certain they are not safe. Even if it takes longer, it is good for her to do them.

Help your child plan what she needs to do. Young children often need more help than older children. For example, when you ask a 6 year-old to clean up her room, explain what you mean. Tell her to:

- Put her toys in the chest.
- Put her clothes in the hamper.
- Pull the covers up over the bed.

By the time she is older, she should know what it means to "clean up her room."

If your child doesn't ask to do things by herself, you may need to push a little. Parents often find it hard to push a child who has an illness. They feel guilty or afraid. Don't let your child's illness get in the way of helping her to grow up. It's OK to let her fail sometimes. That is part of how we all learn what we can do.

No matter what, don't do everything for your child. When she does things for herself, she will feel good about herself.

You are not the only one who may try to protect your child too much. Other family members may feel that she needs special care. Let them know how you want them to treat your child. Ask them to focus on what your child can do, not what she can't do. She doesn't need their "help." She needs their support for growing up.

Support your child to begin to interact directly with the health care provider. Make sure that your child can say what special things she needs to do, like drink more water, take certain daily medicines, not get over tired and visit the doctor regularly. She also should know when she needs to tell you or a teacher that she is not feeling well.

Teach your child age-appropriate ways to take care of herself, i.e., making some of her own decisions but with your close guidance. Encourage hobbies and social activities, including music lessons, Boys and Girls Club, and sports. Give a small allowance and help your child learn about budgeting it and saving. Make sure that your child does some household chores like washing the dishes, keeping her room clean and sweeping. Encourage her to think about her future by asking "what will you do when you grow up?"

Self-Esteem

School Sports: For Boys and Girls

One of the ways that many children build self-esteem is through sports. If your child wants to play school sports, help her to be realistic about what she can do. Talk to your doctor about sports. Look at her strengths and find a sport that matches these.

If the sports program has a leader or coach, tell them about your child's special needs. Remind your child that she needs to take good care of herself. She needs to:
- Drink when she is thirsty.
- Rest when she feels tired.
- Dress for the weather.

Help your child feel good about herself. Pay attention to more than her problems. Notice her skills, her strengths, her interests and her style.

- **Praise her when she does well.** You can't give too much praise. Children thrive when they are told that they are special. They feel good when someone sees something they've done well.

- **Listen to what she has to say.** Ask her questions. Show her that you care about what she thinks and feels.

- **Help her get involved in things besides her illness.** She can try some of these things to see if she wants to do them:
  - Playing music
  - Being with friends
  - Learning the computer
  - Drawing or working with clay
  - Playing chess or other board games
  - Acting in a play
  - Reading books

Be careful not to push your child to compete where she can't succeed. If she can't run very long without getting sore or tired, don't force her to play fast sports. Help her to find at least one thing she does well and enjoys.

Help your child try new things, like learning how to use a computer.

Accepting Herself

When your child starts school, she may begin to notice that she is different from other children. When she becomes aware of having "a disease," she may feel afraid or angry. She may think that she got sick because she did something wrong.

Make sure your child knows that she didn't get the disease because she was "bad." Nothing she did gave her the disease. Nothing she can do will get rid of it.

Your child needs to accept the fact that she has sickle cell disease and make the most of her life. Tell her that except for her disease, she is just like other children. Help her learn what she can and cannot do so she can gain more control.

Fitting In

We all like to feel that we belong. Your child may be afraid that she won't fit in because sickle cell disease makes her different. She may think that others will make fun of her or treat her "special."

Some children feel good about telling their friends about sickle cell disease. Others don't want anyone to know. It is good for children to tell at least one close friend about their disease. If no one knows, it is more likely that they will feel ashamed of their secret. When friends know, they can support your child when she needs it.

If your child doesn't know what to say to her friends, let her practice with you first. Her friends will handle it best if she tells them what the disease means and that they can't catch it from her. Once she has told others, she will probably feel better about herself.

School Success

Most children want to do well in school. School is their work. Doing well in school builds a strong future.

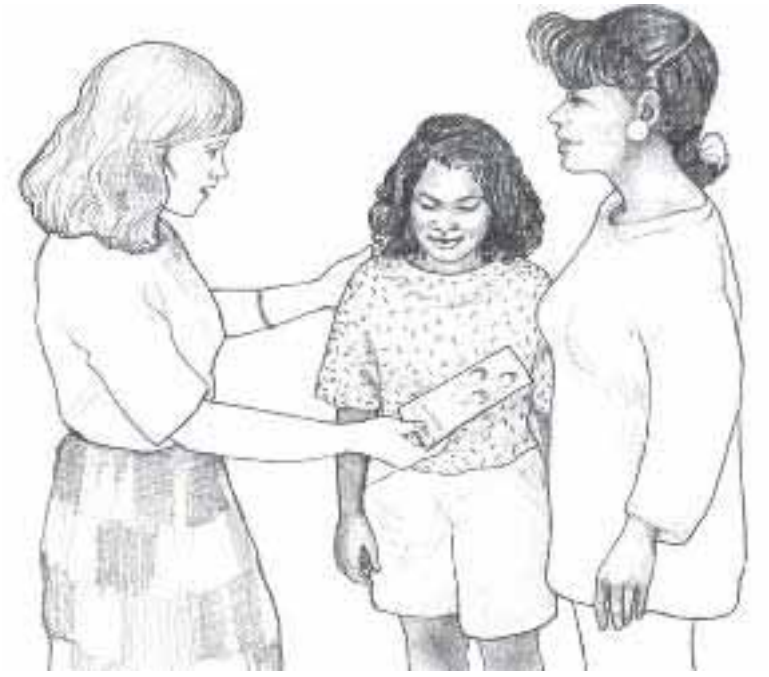
Be a Partner with Teachers

At the start of a new school year, go to school and meet your child's teacher and school nurse. Bring your child with you. Let your child ask any questions that she may have and help her feel at ease. Tell the teacher about sickle cell disease. Give the teacher things to read so she can learn about the disease.

The teacher needs to know that your child will come to school even when she has minor aches and pains. She should be sent home only if she has a fever or severe pain or if she needs to see a doctor.

Explain your child's special needs. She needs to:

- Get water when she is thirsty.
- Go to the bathroom as soon as she feels the need.
- Make up school work if she has to miss school.



Give your child's teacher information about sickle cell disease.

- Rest or slow down if she is tired or sore. For example, during gym class, she may only be able to run 2 laps, not 6.
- Rejoin the class as soon as she is ready.
- Get medicine if she needs it.

Check to see that her teacher gives your child what she needs. Some teachers may protect your child too much while others may ignore her. Talk to the teacher about these things if you are concerned. If you need help or support, talk to your doctor, nurse or social worker.

Stand Up For Your Child's Rights

Your child has the right to get an education that meets her needs. There is a law which says that the school has to give it to her. This law (PL94-142) means that the school must provide help if your child needs it.

If your child is not doing well in school, talk with her teachers. You may want to ask that she be tested for learning problems. If she has a learning problem, she should get special help so she can learn better. Ask the school counselor for an "individualized educational plan (IEP)." This plan could include any of these:

- Regular school classes
- Home instruction
- Time with a resource specialist
- Special classes

If a teacher suggests that your child be held back, get an opinion from another learning expert. Often, being held back is not helpful to children with learning problems. Make sure that your child is tested for these problems and gets any extra help she needs.

Tell your doctor or a social worker if you don't think your child is getting enough help. They can help you work with the teacher and school so your child gets the help she needs.

Plan Ahead for Illness

Set up a plan with your child's teacher for your child to do homework or make up the work that she misses if she gets sick. Talking about the plan will give you a sense of what the teacher thinks about your child's disease. It is a good time to give the teacher more facts about sickle cell disease. Most teachers are happy to do what they can to help.

Find out what resources your school has for children who have a chronic illness. Ask your child's teacher if there is a hospital tutor program or other community programs that could be helpful if needed.

If you don't think that there is enough help, **speak up before your child needs it.** Ask the social worker who works with your sickle cell program, a family member or friend to help you speak up about your child's needs.

The more your child is in school, the better off he'll be.

Keep Your Child in School

Send your child to school unless she is sick enough to see a doctor. Don't keep your child home from school if she just has something like a runny nose. She also doesn't need to stay home because of bad weather. Just make sure she wears the right clothes.

Keeping your child home from school when she doesn't need special care will cause problems. She may be left out of friendships and have trouble learning social skills. She may also find it harder to do well in school.

If your child does have to spend time in the hospital, have her try to do her homework in the hospital. You can also encourage her to talk or write about what has happened to her in the hospital. If she misses much school, she may need outside help or a tutor so she can keep up.

Most of the time, it's best to send your child back to school as soon as she comes home. The more she is in school, the better off she'll be.

Unless your doctor says it is needed, don't agree to home instruction for your child. Home instruction can't replace the school setting. In the classroom, children learn from each other as well as from books and teachers.

Expect the Best

Like other children, children with sickle cell disease can excel in school. Both you and her teachers need to expect the best from your child. When you expect more of your child, she will do better.

Some teachers may not expect enough from your child. Don't let a teacher protect your child from learning what she needs to learn. If your child is not doing well, she should get help.

Care for Yourself and Your Family

Other People Can Help You in Many Ways

Friends, relatives and neighbors can:

- Listen to you.
- Take you and your child to the doctor.
- Spend time with your other children when you have to give your child with sickle cell disease special care.
- Include your child with sickle cell disease in parties, outings and other social events.
- Tell you about places to get help or other resources.
- Comfort you when you're feeling down.

Even though you do all that you can to help your child stay well, she may still have problems. And these problems can affect your whole family.

As your child grows up, you may face new problems, like learning problems or pain. You may feel scared and angry again, even though you thought those feelings had left for good.

It is important for you to take care of yourself, as well as your child. Learn about your limits and needs. Sometimes you may need time for yourself, your other children or your work. Ask for help so you can have the time you need.

Your Other Children

Your other children need your attention and care, too. Make time to talk to them and be with them. Try not to miss school or sports events that they are involved in because you are focused on your child with sickle cell disease.

It can be helpful to teach all of your children about sickle cell disease. If they have questions that you can't answer, let them ask the staff at the sickle cell center. Knowing more about the disease will help them feel included.

One of the keys to a healthy family is to treat your child with sickle cell disease like her brothers and sisters as much as possible. Try to use the same system of discipline and rewards with all of your children. Special treatment isn't good for any of them.

Ask for Help and Support

Chronic illness raises questions and concerns for all families. It can help to have someone to talk to about your concerns.

Family members can be a big source of help. Share your problems and successes with them. Grandparents are often the most helpful. If your family wants to help you out, let them.

You can also get help from your church, social worker or counselor. Or look for a support group with other parents of children with sickle cell disease.

Don't be shy about asking for help. If you hold on to problems for a long time, they can be harder to solve.

See Appendix J for a list of Parent Support Groups in California.

"My son said he wished he had sickle cell disease like his sister. I told him that he was special, too."

The Teen Years

Teens can still have many of the same problems which younger children have from sickle cell disease. Infections, pain, low blood counts and strokes can affect people with sickle cell disease at any age.

Some of these problems may be less frequent for teens, like infections. Most fevers can now be treated with antibiotics at home, not an IV in the hospital. Still, your teen needs to see a doctor for a fever over 101° F.

Here are the most common health problems that teens with sickle cell disease face:

- Pain
- Eye Problems (retinopathy)
- Leg Ulcers
- Avascular Necrosis (hip and shoulder pain)
- Appearance (delayed growth)
- Infections
- Gallstones
- Acute Chest Syndrome
- Priapism

Many of these problems have been described in detail in earlier chapters. Pain and the first four problems will he described in this chapter.

Pain

Some teens with sickle cell disease have more pain as they get older. The pain may feel worse or just come more often. The treatment for pain is the same for teens as it is for children. (See Chapter 3 for more about pain.)

Encourage your teen to take the time to drink fluids and rest.

By now, if your teen has had a problem with pain, he has most likely tried lots of ways to ease it. It makes sense for him to use the ones that work best for him.

Pain Triggers

Dehydration (lack of fluid in the body) is a common trigger for pain at this age. Teens often get involved in sports and don't take the time to stop and drink or rest. They need to take responsibility and find ways to take care of themselves since you can't always be there to remind them.

Rarely, girls get severe menstrual pain which brings on sickle cell pain. The doctor may be able to prescribe a hormone to prevent this pain.

Your teen will learn more about what triggers his pain. As he does new things, suggest that he watch to see what happens with the pain. If he finds that something often leads to pain, he can stay away from it or take special care with it.

Pain Therapy Agreements

For teens with chronic pain, a written agreement may be helpful. The agreement is about how pain will be handled. These agreements are often used in the hospital, but can be helpful at home.

Pain agreements have two parts:

1. The first part says what the health care staff will do to help the teen manage his pain.
2. The second part states what the teen will do for himself to manage the pain.

See Appendix K for a sample Pain Management Agreement

NOTE: If a teen has a fever with pain or if the pain is in his stomach or chest, he should see a doctor. Call first so he can be seen right away.

Eye Problems

Sickle cell disease can cause eye damage and, rarely, blindness. This is why your teen needs to be checked by a special eye doctor (an ophthalmologist) once a year. By the time your teen complains of poor vision, the changes may have gone too far to correct.

The back of the eyes contain tiny blood vessels that can become clogged by sickle cells. This can cause bleeding or scarring at the back of the eyeball. When the damage starts, you can't see it. Only an eye doctor using special techniques can see it. If the doctor finds damage, it can be treated. Without treatment, these early changes can lead to loss of vision.

Leg Ulcers

Ulcers usually start as a small sore on the ankle. They can grow large and get infected. Some ulcers heal quickly, while most take a long time to heal.

Sickling happens more in the lower legs and ankles because of the pressure of standing. With sickling, small blood vessels get blocked, and blood can't get through to all of the cells. An ulcer forms when lack of blood flow to the ankle skin kills the skin cells.

Men are more likely to get ulcers than women. People with SS disease are also more likely to get ulcers than those with other kinds of sickle cell disease.

Put on a fresh bandage every day.

It is much better to treat leg ulcers when they are small than when they are larger. Skin ulcers frequently recur.

Signs of Ulcers

Take your teen to see the doctor if you see either of these signs:
- **A cut or wound that doesn't heal**
- **A patch of dry, itchy skin**
- **A small dark spot that is surrounded by painful swelling.**

Treatment

These are the basics to good treatment for ulcers:
- Keep the area very clean.
- Stay off the feet as much as possible.
- Elevate the feet as much as possible.
- Put on a fresh bandage twice a day or as often as advised.
- Wear clean white cotton socks and flat protective shoes until the ulcer is healed.
- Use lotion or ointment to keep the skin moist.
- If the ulcer looks infected, see your doctor for antibiotics.

If the ulcer is large or has not started to heal in a few weeks, your child may need to go into the hospital. There, he will be on strict bed rest and get special wound care. Transfusions may also be used to try to bring more oxygen to the tissues.

If the ulcer doesn't heal, surgery may be needed. A piece of skin from the thigh may be used to cover the ulcer. This is called a skin graft. It means a longer stay in the hospital for strict bed rest. If it doesn't work, another skin graft may be needed.

Avascular Necrosis (Bone Damage)

This is the term for damage to the bones caused by sickle cell disease. Usually the hips and shoulders are the joints that are affected. When the hip bone is involved, it can cause chronic pain in the hip joint when a person walks.

When the blood flow to the hip joint is slowed by sickle cells, the bone in the joint becomes flat and crooked. Then the hip can't move freely. Walking puts pressure on the joint, and more damage is done.

Your teen will know if he has this problem. His hip will hurt when he walks or runs. He may also have lower back or thigh pain. Report any severe pain in the hip joint, legs, or shoulders to the doctor as soon as possible. The best test to see if this is a problem is magnetic resonance imaging (MRI). Early treatment helps.The treatment depends on the extent of the problem. Sometimes a person needs to use crutches for a few months to take the weight off the joint. Other times, your doctor may suggest physical therapy or surgery to stop the hip from changing shape.

If your teen can't walk without severe pain, the hip may need to be replaced. This can only be done when the bones have stopped growing. If your teen needs this treatment, he will need to wait until he has grown to his full size.

Appearance

Late Puberty

Some teens with sickle cell disease reach puberty about two years later than others their age.

Puberty means many changes. These changes are the same for teens with and without sickle cell disease. The only difference is when they happen. For girls, puberty is when their breasts grow and their periods start. In boys, their face hair grows, their muscles get bigger and their voices deepen. Puberty also means being able to get pregnant or to make someone pregnant.

Late puberty is not a problem in itself. But it can make your teen feel bad about himself. Talk to your doctor or a counselor if you or your teen feel concerned about this issue.

Smaller Size

Some teens may be small and thin for their age. Children with sickle cell disease catch up late in their teen years. Although they may be worried about being short, most people with sickle cell grow to normal height. If your teen is very small or thin, talk to your health care team. Usually, nothing is needed. In a few cases, tests may be done and treatment recommended.

Yellow Eyes

People with sickle cell disease can have yellow eyes from time to time. This is caused by a yellow colored substance called "bilirubin" that comes from broken down red blood cells. In some people, the yellow tint lasts for a long time. It may always be there. It is not a medical problem unless the eyes are much more yellow than you've seen before. If the color is much stronger, call your doctor to see if your teen needs to be checked.

What Can Be Done

If your teen is concerned about his looks, reassure him. You and other family members can support him in feeling better about himself. It may also be helpful for him to talk with a counselor or other teens with sickle cell disease about his feelings.

Living with Your Teen

The teen years are a time of major changes for you and your child. There is a lot that you can do to help your child during these years. There is also a lot that is out of your control. Your support and help still matter, but your teen will be making his own life choices.

The teen years can be hard for families, with or without a chronic illness. There are also special problems for this age group that come with sickle cell disease. These problems can make these years a challenge for everyone.

Your teen will learn to take care of himself more and more during these years. He is getting ready for the time when he will be on his own. He is growing up and learning to live his own life. Your teen needs limits, but he also needs freedom. You and your teen will keep trying to find a balance that works.

As a parent, there is much that you can do to help your teen during these years. Each of these guidelines will be described in this chapter.

- ♦ Let Your Teen Do for Himself
- ♦ Set Limits for Your Teen
- ♦ Build Self-Esteem
- ♦ Help with School and Future Plans
- ♦ Get Help for Your Teen and Your Family

Let Your Teen Do More

Your teen will soon be an adult who will be living on his own. Now is the time for you to help him take control of his life.

It can be hard for a teen with sickle cell anemia to feel in control of his life. Many teens feel chained to the disease and the special care it requires. They feel angry that they have to listen to lots of adults - you, teachers and doctors. While this is true, there are ways that they can have more control over their lives even with their disease.

Starting To Let Go

Your teen may not take care of himself as well as you took care of him. Try to let him do it anyway. If you are afraid that he is hurting himself, talk to him about it. Explain what might happen and find out why he doesn't want to do something. Help him figure out a better way to take care of himself.

Your teen wants to be herself.

Now is the time to shift control from you to your child. You can start slowly, but keep moving towards giving him more control. You need to trust him to ask for help when he needs it. If he makes a mistake, help him learn from it. If he learns now, he can take charge of his own life as an adult.

Doing Other Things His Own Way

There are many ways for your teen to be independent besides caring for his disorder. The way he talks, the clothes he wears, the music he likes, and the way he does his hair are all ways to express himself.

You may not like the way he looks or acts, but try to accept it. Unless he is hurting himself or others, try to leave him alone. He wants to be different from you. He wants to be himself.

Growing Up May Be Scary

Many teens with and without sickle cell disease are afraid to grow up. They don't want to take care of themselves and face their lives. They may try to act younger than their age. A 13 or 14 year old with sickle cell disease may look 10 years old. Since they often look younger, you and others may be tempted to treat them that way. Don't let this happen. Expect your teen to act his age, not his size. Stop doing everything for him. If you don't do it all, he will find out that he needs to take care of himself.

It can also be helpful for your teen to get more involved in things other than his disease. Help him find a part-time job, a hobby or new friends. This will give him a chance to be with others and prepare for his future.

Gaining More Independence

Most people with sickle cell disease can look forward to growing up to become independent adults. As a parent, it is your role to prepare your teen for adulthood and living on his own.

As your teen gets older, you should encourage his independence, no matter what your child's capabilities are. Even if your child has severe learning disabilities, or is in full time special education, he should be encouraged to do some things for himself.

Help your teen identify and build on his strengths. Discuss issues related to reproductive health, risky behaviors that teens can get into and substance use. Talking about it won't push your teen to do these things but he needs your guidance in making the best judgements.

Your teen should know the name of his condition (i.e., sickle cell anemia or SS disease, sickle C disease or SC disease, sickle beta thalassemia disease or S beta thal disease, etc.). He should know what kinds of medicines he is supposed to take every day, at what doses. He should know the steps to take when he is having pain, what triggers his pain and how to prevent pain when he can. He should keep track of his medical appointments and start to make his own appointments, although you should make sure he stays on schedule with appointments.

You can go to appointments with him but he should see the health care provider by himself for part of the appointment. You can still fill his prescriptions and monitor that he is taking his medicines regularly.

Talk with your teen about adult health care options and what things will be the same or different in the adult health care setting. Talk about financial resources, both health care related (i.e., insurance) and related to daily living so that your teen has a good plan about how and when he might be able to support himself or contribute to your household. Encourage work and volunteer activities, talk about and help your teen explore possible career interests.

By the time he is a senior in high school, he should be filling his own prescriptions, making his own appointments and getting to them on his own. Once he is 18 years old, the health care providers cannot talk with you without your child's permission, so make sure he is ready to communicate independently!

Keep important medical records (e.g., immunizations, medical history) together in a folder and give a copy of everything to your child when he goes away to college. Some college bound teens can benefit from asking for special accommodations through student services at the campus.

Groups Who May Offer Help on How to Parent A Teen

- Your local YMCA or YWCA
- Church groups
- Schools
- Local substance abuse prevention programs
- Support groups for parents of teens
- Support groups for parents of children with sickle cell disease
- Parents Anonymous

Sickle cell support groups can be a great help.

Set Limits for Your Teen

As the parent, you are still in charge. You have a right to know what your teen is doing and who your teen is with. It is your job to decide how much freedom to allow.

Limit Setting with Your Teen

Limits provide rules, boundaries and guidelines to protect your teen and support their growth. Limits can cover many issues like curfew, homework, chores and use of the car. Communicate with your teen and tell them how you expect them to act. Make sure that the limits are clear to both of you.

If the rules are broken, you should discuss the action and consequences with your teen. The disease is not an excuse for breaking rules. Your teen needs to be held responsible for his choices.

Sickle Cell Disease and Risk Taking

Like other teens, young people with sickle cell disease sometimes take risks. Many teens have a need to prove that they fit in with their friends. Drugs and alcohol, sex and fast driving are all things that some teens do to prove themselves.

Your teen needs your guidance and attention.

Teens with sickle cell disease may have a stronger need to prove that they fit in. They may also be depressed and want to escape from their pain. This can lead them to take risks that can be hurtful to themselves and others.

Some of these risks may carry extra danger for your teen with sickle cell disease:

- Sex without condoms can bring a greater risk of getting a sexually transmitted disease (STD).
- Getting pregnant can be more of a problem for your teen.
- Alcohol can increase sickling because it dehydrates the body.
- Cigarettes can also increase sickling because smoke lowers the oxygen level in the blood.

All of these are risks for any teen. It is just that they can cause more harm to teens with sickle cell disease.

When to Step In

If your teen is taking risks that could be harmful, you need to get involved. Your teen needs proper and fair discipline if he is hurting others.

He also needs your guidance and attention. Listen to your teen. Find out what is behind his actions. Ask why he is taking risks, and listen to his answers. Tell him how they could hurt him and ask him what he thinks.

If your teen doesn't stop or can't stop taking these risks, he needs help. Taking risks that put him and others in danger can be a sign of a deeper problem. You may need outside support and help.

Most people are not taught how to be parents, let alone parents of teens. It is OK for you to be confused about how to handle your teen. It is OK to get help.

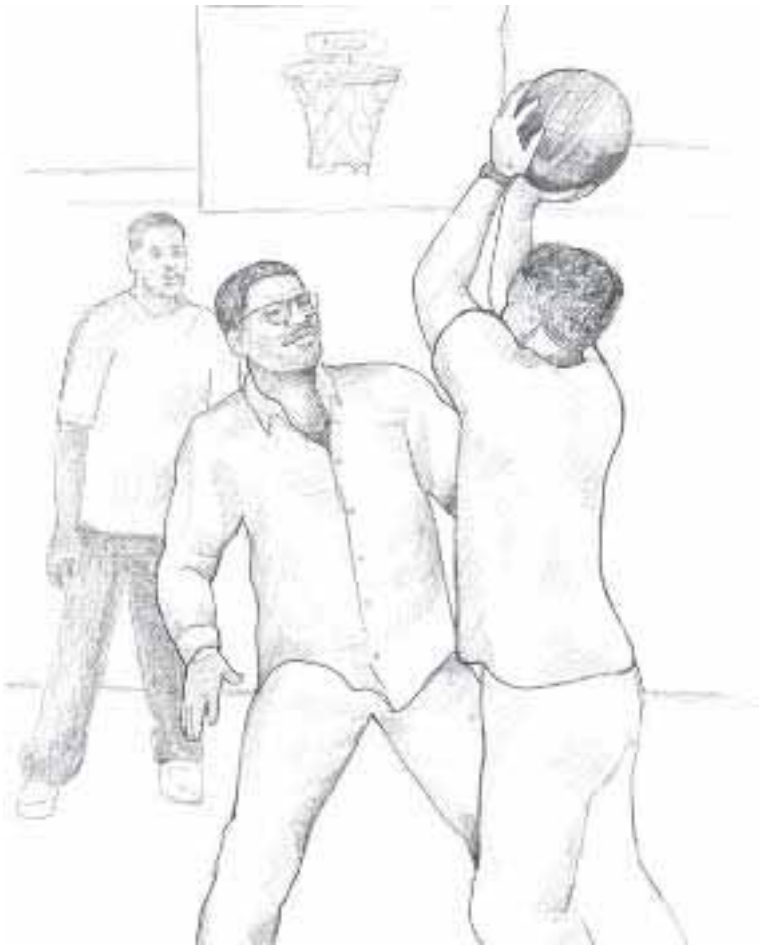
Build Self-Esteem

Like all teens, teens with sickle cell disease may sometimes have low self-esteem. There are many things you can do as a parent to help your teen feel better about himself:

- Listen to what your teen says.
- Don't put your teen down.
- Encourage your teen.
- Do things with your teen.
- Help your teen feel better about his body.
- Let him know how good you feel about him.

Listen To What Your Teen Says

Your teen needs to feel that you listen to him when he speaks to you. Even if you don't like what you hear, you can still try to understand what he is saying. A good way to let him know that you have heard him is to repeat what he has said. Once he knows he's been heard, you can then tell him how you think and feel.



Spend time together doing something you both enjoy.

Ask questions if you aren't sure what your teen means or how he feels. Questions that take more than a "yes" or "no" response are the most helpful.

Don't Put Your Teen Down

It can be easy to put down teens and say hurtful things to them. Put-downs are harmful, not helpful. They won't make your teen want to change or feel good enough about himself to change. Instead, they just make it harder for your teen to relate to you.

If you don't like something that your teen did, sit down with him and talk about how it affects you. Your teen needs your respect and praise.

Doing Things with Your Teen is Important

Being with you may not be your teen's favorite thing to do. But he still wants to know that you want to be with him, at least sometimes. Think about something that you would both enjoy. It could be a movie, a ball game, a concert or anything that you both like to do. Spending time with your teen can tell him that you think he is worth being with.

Your teen may also want you to be involved with his health care sometimes. Check and see how things are going from time to time. Once you've stopped taking care of all your teen's health care needs, it can still be nice to help him out sometimes.

Help Your Teen Feel Better About His Body

Teens with sickle cell disease often feel ashamed of their bodies. Some look younger than their friends because they mature late. Some feel embarrassed by jaundiced eyes. Some have scars from surgeries and IVs. Besides the things that can be seen, some teens may also have limits on what they can do, like sports or dancing. Some teens want to prove to themselves they are like other teenagers and engage in behavior that may not be appropriate.

There are some things you can do to help your teen feel better about his body. If your teen is small for his age, help him find clothes that are right for his age and look good on him. If he has scars, help him find clothes that cover them up. You can also help him find easy ways to talk about his differences to others. If he can talk about them, he may not feel as ashamed.

Remind your teen that his body will mature, and he will get bigger. It will just take him a few more years than his friends. Also let him know that it is OK to be his current size. Focus on his strengths and help him feel good about them.

Encourage and Praise Your Teen

We all need praise. Providing your teen with regular praise encourages success.

Keep an eye out for the times your teen does good things for himself and for others. Don't be afraid to tell him how good it makes you feel when you see him doing things well. The more good things you notice, the more good things there will be.

Don't assume your teen knows that you love and care about him. Teens need to be told that they are loved and that they count.

Treat your teen like he has something to offer, and he will. Help him get involved in things that he does well. Ask him about school, work or any special projects. Ask him to show you what he is working on. Your attention will show that you believe that what he does has value.

Praise your teen for a job well done.

Help with School and Future Plans

Staying in School

Many teens with sickle cell disease have missed a lot of school because of their illness. Some of these teens have managed to do well in school. Others have major problems. They may be behind their classmates, have poor study habits, or overlooked learning problems.

If your child is having problems with school, these problems may get worse during the teen years. He may feel depressed about his future. These hopeless feelings can make it hard to succeed. If your teen feels like he can't keep up, he may just want to give up and drop out.

Urge your teen to stay in school. Help him keep up his drive to do his best. Encourage him to focus on what he wants to do in the future and to work towards it.

Your teen can arrange to make up school work he misses if he has to be out of school. Also, find out if the school has help for students with special needs. Help your teen get extra help if he needs it.

Planning for the Future

High school is the time for your teen to focus on his goals for the future. Help him focus on what he can do, not what he can't do. No matter what his limits, there is a place for him. As he starts to plan his future, help him look at his strengths and skills and see what type of work might be good for him.

It may help him to have a model of success. If you know an adult with sickle cell disease whose life he might admire, tell him about what that person is doing. He might even want to meet him or her.

A part-time job can be a good way for him to learn more about his interests. A job can also help your teen gain confidence in his skills and earn some money. During the school year, make sure that the job doesn't get in the way of school. During summers, a job can be a great way to help teens mature.

Your teen will need to decide whether he wants to go on to college. Many people with sickle cell disease go to college. Some colleges have programs to help students with special needs.

Encourage your teen to plan for a full life. When he takes some control of his life, his image of himself will improve, and he will have a better chance for success.

A job can help a teen gain confidence in her skills.

Job Worries

Many teens with sickle cell disease worry about whether they will he able to find a job. They are afraid that they won't he able to support themselves. They may feel scared that their limits will prevent them from being able to work or keep a job.

Listen to his fears and give him support. Tell him that some people with sickle cell disease have become doctors, lawyers, social workers, teachers and business owners. Help him focus his mind on what he can do to get ready for his future.

- When you allow him to take care of himself, you are preparing him for the future.
- When you help him feel better about himself, you are raising his chances of success.
- When you help him do well in school as well as get involved in other interests, you are giving him the best training for future work.

Places for Teens to Get Help With School or Work

- School counselor
- Vocational counseling
- Educational consultants
- Tutoring programs
- Guidance clinics
- Churches
- YMCA or YWCA
- Social worker

Help for Teens

If your teen is having problems with school or doesn't have plans for his future, he needs help. A counselor from the school or social worker from the clinic can help him take a look at his feelings and his life. The counselor/social worker can work with him and help him find better ways to live with his illness.

Help your teen do her best.

Get Help for Your Teen and Your Family

Get Help for Your Teen

There are times when your teen may need more help than you or his health care team can provide. These are some signs that your teen needs help:

- Your teen is angry a lot or taking dangerous risks.
- Your teen seems very depressed or anxious.
- Your teen won't take care of his health.
- Your teen is doing poorly in school.
- Your teen has more frequent visits to the emergency department and/or hospital.

Get More Help from Your Family

Families often have tough times with teens. When any of these things happen, reach out for help for your family:

- You and your teen feel like you can't talk to each other.
- Your other children are very upset about the way your teen is acting.
- Someone close to your teen gets divorced or dies.

Get Help For Yourself

When teens have problems, parents have problems. Getting help for yourself is often the best way to help your teen. With support and guidance, you will be better able to give your teen what he needs.

Get Help Before Problems Get Worse

Getting help early is a sign of strength, not weakness. It is an important way for you to help your teen and your family make the most of your lives.

Where to Go For Help

Find a counselor or social worker who knows about sickle cell disease and chronic illness in teens. Most sickle cell centers have social workers and counselors on their staff who know a lot about the disease. Or ask your doctor or nurse who they think would be most helpful to your teen or your family. You may also be able to get help through your church.

Many cities have support groups for parents of children with sickle cell disease. Some large cities also have support groups for teens with sickle cell disease. Ask your doctor if there is a program like these that you or your teen can join. Parents and teens can gain a lot from talking with other people like themselves.

See Appendix J for a list of Parent Support Groups in California. See Appendix L for a list of Sickle Cell Centers in California.

Find a counselor who knows about sickle cell disease.

Sex and Reproduction

In the teenage years and through adulthood, people face choices about sex and having children. It is not always easy to know what is right to do. This is true for both children and parents.

People with sickle cell disease face the same challenges as other people. They need to decide when they want to have sex. They also need to choose if they want to have children. There are some differences in how these choices may affect their health. These differences will be discussed in this chapter.

Parents can help their children face these important questions. You can talk to your children so that they know the facts. You can also share your feelings and values to help guide their choices. This chapter will help you talk to your child with sickle cell disease about these issues. It will also inform you about the impact of the disease on these areas of your child's life.

This chapter will address:

- ♦ Talking to Your Teen About Sex
- ♦ Birth Control Basics
- ♦ Sexually Transmitted Diseases and AIDS
- ♦ Pregnancy

Sex and Teens

Like all teens, teens with sickle cell disease face challenges with sex. As they reach puberty, their bodies go through many changes. They begin to have strong sexual feelings. Girls are able to get pregnant, and boys are able to make someone pregnant. All teens have choices to make which will have a major impact on their lives.

Questions for Teens To Answer

Here are some of the questions which teens need to answer for themselves:

- How will they respond to their sexual feelings?
- When will they have sex? With whom? Will they wait to have sex until they are married?
- Do they want to have children?
- If they have sex but don't want children now, what kind of birth control will they use?
- How will they protect themselves from the AIDS virus? What about other STDs (sexually transmitted diseases) like chlamydia or genital warts?

These are not easy questions. Teens need to give these questions a lot of thought and make good choices for themselves and their future families.

Listen to your teen and share your values.

Concerns With Sickle Cell Disease

Teens with sickle cell disease may have concerns about maturing late. Boys may wonder whether they can have sex if they have not reached puberty or if they have a problem with priapism. Boys and girls may worry about whether they will be able to have children.

These concerns can add to normal fears about dating. Teens with sickle cell disease often worry about whether they will find a boyfriend or girlfriend. Having a good time with other teens of both sexes may ease some of these worries.

Teens need to be reassured that they will be able to get pregnant and have sex like other people. Remind them that getting pregnant or having sex are not good ways to prove that they are OK.

Talk To Your Teen About Sex

You don't have complete control over what your teen does about sex, but you can talk to her. You can give her the facts and help her make sense of all that she has heard. You can share your feelings and values so that she knows where you stand. You can listen to her feelings and thoughts and help her decide what is best for her.

Don't wait until your child becomes a teen to start talking about sex. Your child may ask you questions about these issues. Answer what she asks. You don't have to explain everything at once. If your child doesn't ask you about sex, bring up the subject. Look for times to ask about her thoughts or feelings and to share yours. It could be while watching TV or when you hear or read something about sex or love.

Don't assume your teen knows it all even if she acts like she does. Research shows that teens who know the **least** about sex are more likely to have sex sooner. More often than not, it is what teens **don't know** about sex that can hurt them.

Birth Control Basics

People with sickle cell disease can have children. Girls who have reached puberty can get pregnant. Boys past puberty can get a girl pregnant. If they choose to have sex and don't want children, they need to use birth control. There are many kinds of birth control available. Only the IUD (intrauterine device) should not be used by women with sickle cell disease.

Condoms and Foam

One of the best forms of birth control for people with sickle cell disease is the condom. Condoms are good because they also help protect a person from the AIDS virus and other STDs.

Condoms don't always keep a woman from getting pregnant. For more protection, many women also use a special foam with a condom. This foam is put in the vagina before sex. It kills sperm that the condom doesn't stop.

The Pill

Most **low-dose** pills are safe for women with sickle cell disease. These pills almost always keep a woman from getting pregnant, but they don't give protection from STDs (sexually transmitted diseases) or the AIDS virus. Some couples use both condoms and pills.

A prescription is needed to get the pill. Once the pill is started, it has to be taken every day.

There are many choices of birth control methods.

Depo-Provera (DMPA)

Depo-Provera is a shot given every 12 weeks. The hormone in it keeps women from getting pregnant. It does not prevent STDs or the AIDS virus. Women should ask their doctor about whether it would be a good method for them. Women using Depo-Provera should have bone density studies and may need calcium supplements.

No IUDs!

The IUD is not a good method for women with sickle cell disease. It can lead to pelvic infections which can cause many problems.

Choosing a Birth Control Method

There are other choices besides condoms and foam and the low-dose pill. A woman can also use a diaphragm, cervical cap or other newer methods as they become available.

Your teen should talk with her partner and her health care team about what method of birth control to use. Then she can choose what will work best for her.

Using Birth Control

Planned pregnancies are very important with sickle cell disease. Without planning and special medical care, pregnancy can be dangerous for the mother and the baby. If your teen is going to have sex and she isn't ready for children, she needs to use birth control.

STDs (Sexually Transmitted Diseases)

For More Information

If you or your teen need more information about AIDS or other STDs, ask your doctor. You can also get help by calling one of these **toll free** numbers.

National AIDS hotline: 1-800-342-AIDS

National STD hotline: 1-800-227-8922

STDs are a group of diseases that are spread by having sex. They include diseases like genital warts, chlamydia, and gonorrhea.

People with sickle cell disease are not able to fight infections as well as other people. So if they have sex with someone who has an STD, they are more likely to get one.

Signs of STDs

Most STDs can be cured if they are treated early. If your teen is having sex, she should watch for these signs. If she finds any of these, she should call the doctor right away.

In men:
- Drip from the penis
- Pain or burning when passing urine (peeing)
- Sores, rashes or growths on or near the genitals

In women:
- Strange discharge from the vagina
- Pain in the lower abdomen and fever
- Sores, rashes or growths on or near the genitals

Talk to your teen about STDs.

Some women never develop symptoms even though they may have an STD. Because of this, it is very important that sexually active girls have a yearly gynecology visit. Early treatment can prevent some of the more harmful effects of these diseases.

Your teen should be checked by the doctor right away if she is worried about any of these signs. STDs can make her very sick or infertile if they are not treated.

Talk to your teen about STDs. Explain what they are and how they are spread. If she doesn't have sex, she won't get an STD. If she chooses to have sex, condoms will help protect her and her partner.

HIV and AIDS

Like other STDs, HIV and AIDS can also be spread through sex. Once you have this virus, you can't get rid of it. Over time, HIV can cause people to get sick with AIDS. Some people get sick quickly. Other people stay well for years. As of now, most people who have HIV end up with AIDS.

AIDS is a deadly disease. There are treatments to strengthen the body's immune response and to deal with some of the problems that can occur but there is no cure.

The best way to deal with AIDS is to protect yourself from infection. The main way that people get the AIDS virus is by having sex with someone who has it. Some people also get AIDS by sharing needles through IV drug use.

Talk to your teen about how she can protect herself from this deadly disease. If she has sex or uses IV drugs, she is at risk of getting infected. Go over these guidelines with her:

- **The only sure way to avoid the virus is not to have sex or use IV drugs.**
- **If you have sex, condoms will help protect you from the virus. Also use foam or cream with Nonoxynol-9 to help protect you from the virus.**

In the past, some people have gotten the virus from blood transfusions. The risk now is very low because all blood is carefully checked.

Pregnancy

While a woman with sickle cell disease can have a healthy baby, there are risks. Both she and her baby need to be watched closely. It takes planning and work to have the best chance for a healthy baby.

Women with sickle cell disease may wonder if they can get pregnant and have healthy children. They can. But they need early care to prevent or lessen problems.

Early prenatal care helps the baby. Early care can lower the risk of miscarriage. It can also decrease the risk of having a baby that is too small.

Early prenatal care also helps the mother. The mother needs to be monitored so that any problems can be found and treated early.

Prenatal care should be done by an obstetrician (OB) who is an expert in high risk pregnancy. It is best if the OB knows a lot about sickle cell disease.

A woman with sickle cell disease needs to plan when she wants to have children. From the start of her pregnancy, she has to be careful about what she does. Alcohol, certain medicines and other drugs can all harm her baby. This can be hard for women who rely on certain drugs to help them manage their disease. They may have to live with more pain while they are pregnant or go without treatment for certain problems.

A pregnant woman needs to check with her doctor before taking any medicine. Her doctor will tell her what medicines might cause problems.

It is best to start prenatal care before getting pregnant or right after. The OB and her sickle cell doctor can work together to help keep the mother and her baby healthy.

Some women who are pregnant may need to receive blood transfusions near the last 3 months of pregnancy. This decision is best made with both the OB and the sickle cell physician.

Partners Should be Tested

Both men and women with sickle cell disease can pass the disease on to their children if their partner has a hemoglobin trait. If a partner has a hemoglobin trait, there is a 50% (1 out of 2) chance that a baby will have the disease. Each time they get pregnant, they have the same chance.

A partner should have hemoglobin testing before a couple gets pregnant. Then the couple will know ahead of time if they might have a baby with a hemoglobin disease. The test can also be done when a woman finds out she is pregnant. Talk to a genetic counselor and your doctor to discuss your particular situation.

One Parent Has Sickle Cell **Disease** and
One Parent Has Sickle Cell **Trait**.

When one parent has sickle cell disease and the other has a trait, they have a 50% chance (or 1 out of 2) of having a baby with sickle cell disease. Each time they get pregnant, they have the same chance. (See figures 1 and 2.)

Figure 1:

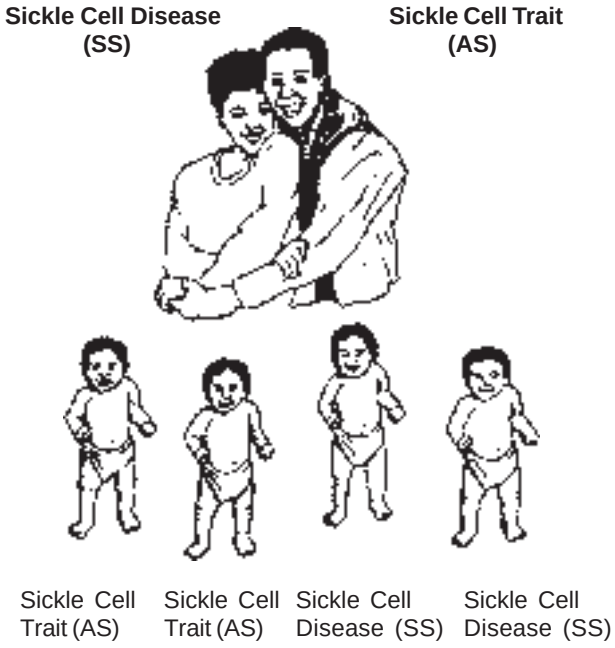
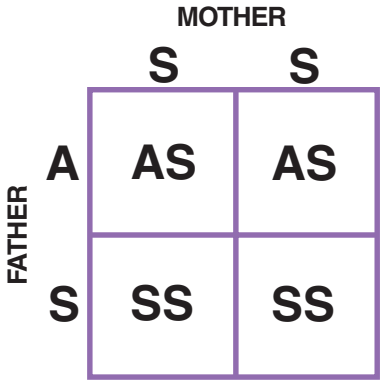


Figure 2:

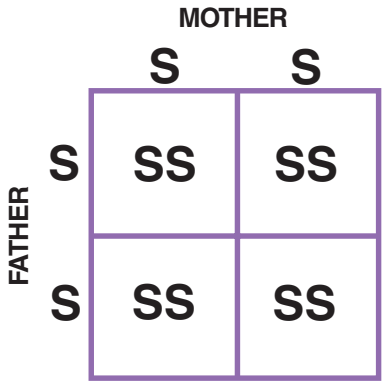


Both Parents Have Sickle Cell **Disease**

If both parents have sickle cell disease, all their children will also have the disease. (See figures 1 and 2.)

Figure 1:

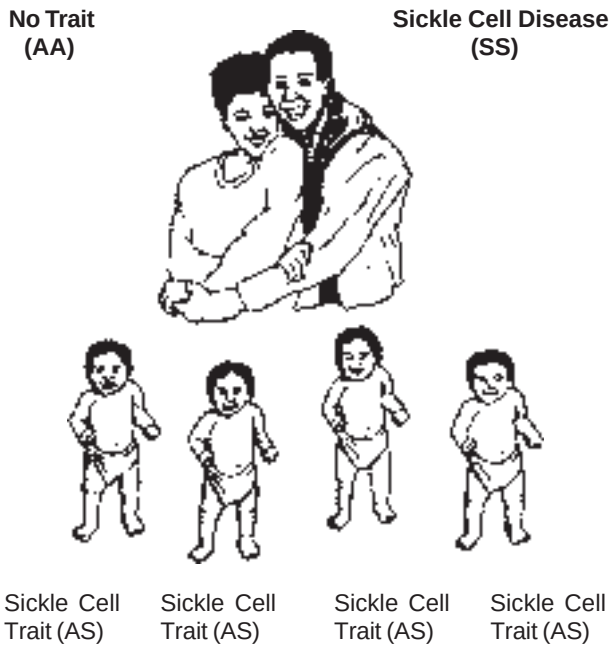
Figure 2:



One Parent has Sickle Cell **Disease** and
One Parent Has **No Hemoglobin Trait**

If one parent has sickle cell disease and the other does not have sickle cell disease or a hemoglobin trait, all of their children will have sickle cell trait. (See figures 1 and 2.)

Figure 1:



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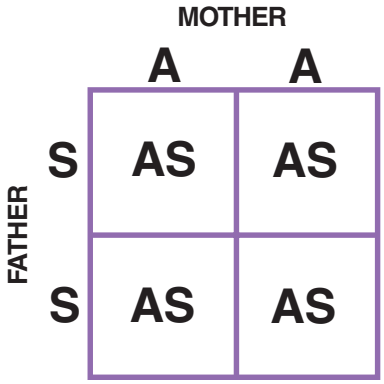


Testing the Baby for Sickle Cell Disease

Many couples want to find out before birth if their baby has sickle cell disease. The unborn baby can be tested in the early months of pregnancy.

If tests show that the baby has sickle cell disease, the couple can make choices. Some people use the time to prepare for their baby's special needs. Others choose not to continue the pregnancy. It is up to the couple to decide what is right for them. The health care staff will support their decision.

NOTE: Some women with sickle cell disease choose not to have children. Others choose to have children. The choice is up to each woman.

See Appendix N for charts to fill in your and your partner's hemoglobin types. Your genetic counselor or doctor can explain how this applies to your family.

Teen Pregnancy

Teens who get pregnant have more problems than older women who get pregnant. Pregnancy is stressful. Since teens are still growing themselves, pregnancy places an added strain on their bodies.

Teens with sickle cell disease are at risk for even more problems. They have all of the risks that teens have plus the risks that come with sickle cell disease. These risks include the chance of having a child with sickle cell disease.

Some teens with sickle cell disease try to get pregnant to prove that they are normal. Talk to your daughter about the risks before she does something that may not be right for her. Reassure her that she will be able to get pregnant when she plans it. Tell her that she does not have to prove that she is fertile now.

If your teen gets pregnant, she will need your support. If she doesn't want the baby or didn't plan to have children, she may be upset or scared. Talk to her. Listen to her feelings and let her know that you will help her. Share your feelings with her, too, but try to do it in a way that doesn't turn her away. It won't help if you stop talking to each other.

Pregnant teens have some hard choices to make. Some teens choose not to continue the pregnancy. Others choose to have their babies. They may raise them or give them up for adoption. No matter what their choice is, teens with sickle cell disease need to see both an OB and their sickle cell doctor if they get pregnant.

Some teenage boys may want to prove that they can father children. Make sure your teenage son knows that he is fertile. If he has sex with a woman and doesn't want her to get pregnant, they need to use birth control.

Where to get help for pregnant teens

• **Obstetrician (OB)**
It is helpful to find an OB who is an expert in high risk pregnancies.

• **Local Health Department**
They may have a public health nurse or social worker who helps teens get the care they need. They may also have prenatal clinic and/or classes for teens.

• **Family Planning Clinics**
These clinics offer low cost pregnancy tests and family planning services. Some of them also provide prenatal care.

• **Community Health Centers**
These clinics often offer low cost pregnancy tests and prenatal care.

• **Medicaid (Medi-Cal)**
This government program pays most or all prenatal care costs for low income women.

• **WIC**
This program provides free food and formula to low income women who are pregnant or mothers of young children.

Your Child's Future: The Adult Years

Work, marriage and family are the main issues of adult life. These are also the main issues for adults with sickle cell disease.

Most adults with sickle cell disease lead full lives. They are able to live on their own and work, take care of a home and raise children. They live with the disease and enjoy their lives.

As a parent, you can help your adult child learn to live on his own. Even adults with serious problems can often live on their own with the help of special programs.

This chapter focuses on two aspects:

- ♦ Health Issues
- ♦ Help and Support

Health Issues

Most people with sickle cell disease can expect to live into middle or late adulthood. As more is learned about this disease, people are living longer.

Find Problems Early

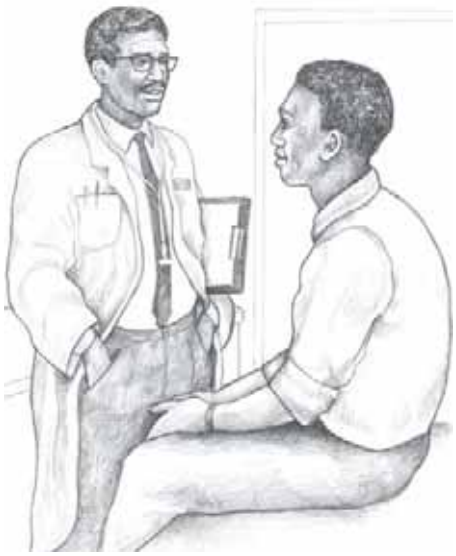
Adults with sickle cell disease still need to see a doctor at least once a year. At these routine visits, their organs are checked for signs of damage. Over time, sickle cells can affect most body organs. The kidneys, lungs, bones, eyes, heart or brain can be damaged by the disease. If signs of damage are found, proper treatment can be started early.

Hospital Care

Most of the health care needs of adults with sickle cell disease can be handled at the doctor's office and at home. Hospital care is sometimes needed.

Major Health Problems

Some adults with sickle cell disease have major health problems. The most common are pulmonary hypertension, chronic lung disease and kidney failure. Other problems, such as avascular necrosis (bone damage), strokes, blindness and congestive heart failure can happen to adults.



Adults with sickle cell disease need to see a doctor once a year.

- **Lungs**
Adults who keep having acute chest syndrome may need frequent blood transfusions. They may also need to have their lung function checked closely.

- **Kidneys**
Some people develop kidney failure and need dialysis or transplants. Transplants can cure kidney disease in many patients.

- **Bones**
The chance of getting avascular necrosis in the hip increases with age. Early identification and care, and sometimes hip replacements, help many people who have this problem.

- **Brain**
Strokes can occur at any age in sickle cell disease. The incidence has decreased since screening for strokes with TCD (see Appendix D) has been available. Transfusion and rehabilitation help most patients regain their abilities.

- **Eyes**
Blindness rarely happens. It can be prevented by yearly check-ups with an ophthalmologist (eye doctor) who knows about sickle cell disease.

- **Heart**
Older adults with sickle cell disease may develop congestive heart failure or pulmonary hypertension affecting the heart. Medicine and transfusions may help.

NOTE: To help prevent and treat these problems, regular visits to the doctor are a must.

Quality of Life

Life with sickle cell disease is not just about what is happening in the body, but also is about what sickle cell disease means to the affected person and his family. On the yearly visit to a sickle cell center, you should be asked about your child's quality of life, or you can go through this inventory annually on your own.

How does sickle cell disease affect how well your child gets around, his behavior and mental health? How does he feel about his health compared with his health one year ago? How healthy does he feel he really is? How much is pain a problem on a daily basis? How is he functioning in school or on the job? How is sickle cell disease affecting other family members emotionally, and in terms of how everyone is getting along and is supporting one another? Are family members able to be involved with usual activities? What does coping with sickle cell disease mean spiritually - are family members despairing or angry, still stuck in "why us"?

Some people have a very good quality of life based on the answers to these questions, no matter how many physical symptoms they have. Others, with just a few physical symptoms, have a very poor quality of life, because sickle cell disease might be the entire focus. If the answers to this yearly inventory about quality of life show that there is room for a lot of improvement, talk about it with your medical team, a counselor or a clergy member.

Fears of Dying

People with sickle cell disease often have fears of dying. A long or serious illness can make these fears worse. These fears can come up over and over again because each new set of problems brings new worries. The same is true for parents. They are often afraid that their children with sickle cell disease will die.

Talk about your feelings with each other. It will bring you closer together, talking about these fears helps most people live with them. Honest, open talk seems to work best. You may also feel better if you share your feelings with others who are close to you.

It is important that the fear of death does not become the main focus of your life or your child's life. If either of you can't stop thinking about it, get help. Talk about it with your medical team, a counselor or clergy member.

Help and Support

There are some programs which provide help and support for adults with sickle cell disease. There are also other sources of support. Both kinds of support are important for adults with sickle cell disease.

Friends, family and others who care can often be a major source of help. Their support is a gift of love that tells a person that he matters.

Formal programs provide a different kind of support. They offer money or other types of help that most people can't afford to give. If your adult child needs these programs, he should make sure he gets them. Some of these programs are described here. Don't figure it out on your own. Talk to the social worker or other clinic staff to figure out where to get help.

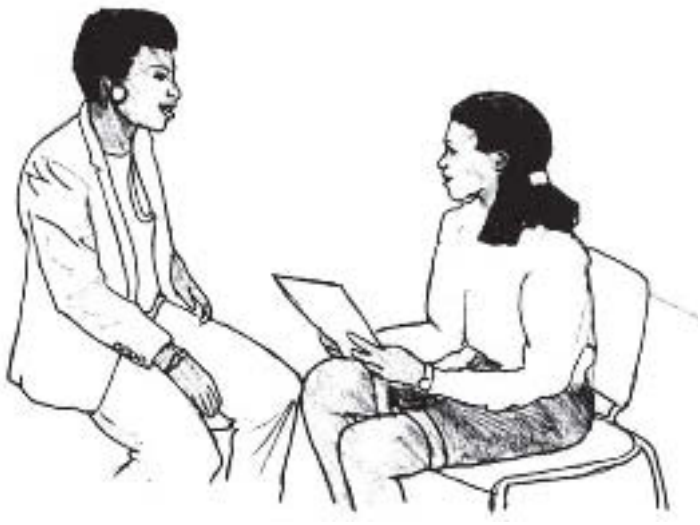
Health Care Costs

Many cities and states have programs to help people who have certain illnesses pay for health care. These programs often cover adults with sickle cell disease.

Job Training and Placement

Adults with sickle cell disease sometimes need help finding a job. Many states have programs to help people with chronic illnesses deal with job and career issues. Ask about job training, career counseling, placement and job support programs.

The Department of Rehabilitation (DOR) assists individuals with disabilities get and keep jobs and maximize their ability to live independently in their communities. Working with people of every type and category of disability, DOR provides vocational rehabilitation services to eligible Californians.



Talk with clinic staff about where to get help.

Vocational rehabilitation services are designed to get Californians with disabilities prepared for employment and can include training, education, transportation, and job placement.

Some of the services provided by the DOR may include, but are not limited to:

- Counseling and guidance.
- Referrals and help getting services from other agencies.
- Job search and placement assistance.
- Vocational and other training services.
- Diagnosis and treatment of physical and mental impairments.
- Transportation if needed.
- Services to family.
- On-the-job or personal assistance services.

Website: www.dor.ca.gov

Basic Living Costs

Some people with sickle cell disease can't support themselves through work. They are fully disabled. The government has a program to help disabled people pay for their basic living expenses. It is called SSI (Supplemental Security Income).

Vocational and other training services are available through the Department of Rehabilitation.

See:
Appendix J - California Parent Support Groups

Appendix P - Resources

Legal Help

Your adult child may have problems getting housing or finding work because of his illness. It is against the law for someone to refuse to hire or rent to people because they have a disease. If your adult child thinks he has not been treated fairly, he should get legal help.

There are legal groups which help people when they have been wronged. Look for these groups:

- Legal Aid
- The Urban League
- The ACLU (American Civil Liberties Union)

These groups have funding to cover the cost of legal help.

Find Out About Help

There are a few places you or your child can go to find out about help. Ask the social worker at your nearest sickle cell center about these programs. Your local Department of Social Services may also know about some support programs.

Many cities also have a local branch of the Sickle Cell Disease Association of America. People in these groups often know the most about the programs in their city. If you have a group nearby, join it.

Support and encourage your adult child in speaking up and asking for help. Encourage him to find out about programs and resources and to find ways to get what he needs.

Advocacy and Self Advocacy

People with sickle cell disease and their families should know about their:

- Disease
- Rights as a patient and family member
- Responsibilities
- Rights as students and employees

People with sickle cell disease and their families need to learn to communicate effectively with:

- Care providers
- Employers and Schools
- Insurance companies
- Legislators

Live with sickle cell disease, don't just survive with it! Know yourself and your strong points (and your child's). Play those up! Act, don't react!

Places to Ask About Help

- Nearest sickle cell center
- Local branch of the Sickle Cell Disease Association of Anemia
- Other sickle cell organizations
- Sickle cell support groups
- Local department of social services
- Local health department
- Local churches
- The Urban League
- Local department of employment services
- Community colleges
- Career planning offices of colleges and universities

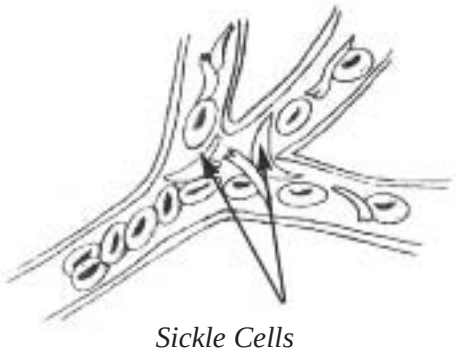
Appendices

What is Sickle Cell Disease?

Sickle cell disease is a disease that affects a special protein inside our red blood cells called hemoglobin. Red blood cells have an important job. They pick up oxygen from the lungs and take it to every part of the body. It is the hemoglobin in these cells that carry the oxygen to different parts of the body.

A person with sickle cell disease makes a different kind of hemoglobin. This causes the red blood cells to change their shape. Instead of being smooth and round, the donut-shaped cells become hard and sticky. Their shape looks like a banana or like a sickle, a hand tool used to cut wheat or tall grass. It is this sickle shape of the red blood cells that gives "sickle cell" disease its name.

The hard, sticky sickle red blood cells have trouble moving through small blood vessels. Sometimes they clog up these blood vessels so that blood can't bring oxygen to the tissues. This can cause pain or damage to these areas.



Sickle Cells

Hard, sticky sickle cells clogging up a small blood vessel.

Normal red blood cells.

Sickle red blood cells.

Types of Sickle Cell Disease

There are many different types of sickle cell disease.

The most common types are:

- sickle cell anemia (SS disease)
- sickle C disease (SC disease)

Sickle beta thalassemia disease (S beta thal disease) is another type, but it is less common.

There are many rare combinations of sickle hemoglobin with other types of hemoglobin. Some are as serious as sickle cell anemia. Some types of sickle cell disease cause fewer problems than others.

What Causes Sickle Cell Disease?

Sickle cell disease is inherited

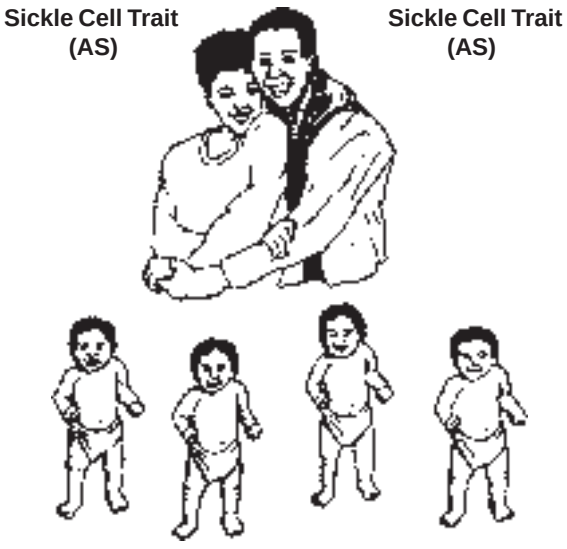
Sickle cell disease is an inherited disease. This means that it is passed from parents to their children through their genes.

Genes are our body's map for development. We have pairs of genes for the color of our eyes, for our height, for our blood type and for each of our other features, including our hemoglobin type.

A baby gets one hemoglobin gene from each parent

To make up our pair of hemoglobin genes, we get one gene from our father and one gene from our mother. Each of our parents has two genes for hemoglobin, but they pass only one of these genes on to each child. Which gene is passed on is a matter of chance, like having a boy or a girl or like tossing a coin and getting heads or tails.

Both Parents Have Sickle Cell **Trait**



To inherit sickle cell disease, a child must get the sickle (S) gene from one parent and a sickle (S), C, or B (beta thal) gene from the other parent. If a baby inherits at least one hemoglobin A (usual adult hemoglobin) gene, he won't have sickle cell disease. (See Appendix M.)

Both parents have a hemoglobin trait

Someone who has one gene for hemoglobin A and one gene for a different type of hemoglobin has a hemoglobin **trait**. This trait could be AS (sickle cell trait), AC (C trait) or AB (Beta thal trait). In addition, there are other less common traits.

A trait is not a disease

People with a hemoglobin trait are healthy. They do not have a mild case of the disease. They do not have a "trace" of the disease. It never changes into sickle cell disease.

How to find out about your hemoglobin genes

The only way to know for certain what type of hemoglobin you have is to have a special blood test called hemoglobin electrophoresis with a complete blood count (CBC). Many families carry genes to make other types of hemoglobin besides hemoglobin A without knowing it. Your doctor or sickle cell center can order this test for you.

NOTE: Hemoglobin type and blood type are not the same. Everyone has both a hemoglobin type and a blood type.

Comprehensive Sickle Cell Disease Care Plan:

6 Years to Adult

Evaluation	Interval
General Physical Exam	
6 - 8	Every 3-4 months
9 - 18	Every 4-6 months
Over 18	Every 6-12 months
Diphtheria and Tetanus Booster	Every 10 years after initial series
Other Immunizations	As advised by your sickle cell doctor
Flu Shot	Once a year
TB Tests	Once a year
Comprehensive Social Worker Evaluation	
Interview, Evaluation, & School Assessment	Once a year
Genetic Counseling Services	
Family Studies	First visit
Sickle Cell Counseling	Once a year or as needed
Hematology Evaluation	Every 4 months to twice a year
Liver-Gall Bladder Evaluation	As advised by your sickle cell doctor
Renal (Kidney tests)	At least once a year
Echocardiogram (Heart tests)	By 10 years with follow-up as indicated
Ophthalmology (Eye tests)	Once a year (after age 10)
Pulmonary Function Testing (Lung tests)	By 10 years with follow-up as indicated
Dental Evaluation	Once a year
Neurological Evaluation	Once a year
Psychological/Family Therapy Consultation	Once a year
Physical Therapy Assessment	As necessary
Formal Nutrition Assessment	Every 2 years or as needed
Sickle Cell Patient Education	1-3 times a year or as needed
Transition Evaluation	Age 12 then yearly
School Performance Evaluation	Once a year

Note: This is the Care Plan recommended by Children's Hospital & Research Center at Oakland Sickle Cell Center. If you have any questions, ask your child's doctor or sickle cell center.

Common Medical Tests

Blood Tests

Hemoglobin electrophoresis
This is the test which is used to find out a person's hemoglobin type. It is this test which tells you what type of sickle cell disease your child has.

This test is also done when it is important to know how much sickle hemoglobin is in your child's blood. Before a blood transfusion, doctors use the test to help decide how much blood should be given. After a transfusion, the test is used to see if enough blood was given to lower the amount of sickle hemoglobin and prevent complications from sickling.

Complete blood count (CBC)
The most common blood test is the complete blood count (CBC). It is mainly done to find out the number, shape and size of the blood cells and the hemoglobin level. This information is used to tell if any treatment is needed.

The normal hemoglobin level in children without sickle cell disease is 11-14. Children with sickle cell disease usually have a lower hemoglobin level of 6-10. This varies with the type of sickle cell disease. If your child's hemoglobin level is less than 6, she may need to be given blood or go to the hospital.

Reticulocyte (Retic) count
Reticulocytes are young red blood cells. The number of these cells shows whether the bone marrow is doing its job well, making and releasing young cells into the blood.

Kidney and liver function tests
These tests show if these organs have been damaged by sickle cell disease. In young children, damage is rare. However, over time, the sickle cells can plug up the small blood vessels of these organs so that they can become damaged.

Other Tests

Bone Density
This test is sometimes called a DEXA Scan; it determines the amount of calcium in the bones. Many people with sickle cell disease have a low bone density and need to take calcium tablets. All pre-teens should have this test to make sure their bones are strong and determine whether they need to take calcium.

Electrocardiogram (EKG)
A test that measures the electrical activity of the heart and gives the doctor information on how the heart is functioning.

Echocardiogram (Echo)
A test that uses sound waves to show an image of the heart on a video screen. It shows a picture of how the heart is working.

Holter Monitor
This is a special test that monitors the heart for twenty-four hours to determine if there are any abnormal beats.

Magnetic Resonance Imaging (MRI)
This is a test that can make pictures of the brain and the blood vessels of the brain. It uses a magnet, not x-rays.

Pulmonary Function Testing
A test of how well the lungs are working. It is very important to have this testing done if you have had acute chest syndrome or if you have had to take medicine for breathing problems. It will help the doctor with your treatment.

Transcranial Doppler (TCD)
An ultrasound used to measure blood flow in the blood vessels of the brain. This is a non-invasive test for stroke.

Ultrasound
Sound waves are used to look at structures in the body. The images are not as clear as with an MRI.

Urine Test (Urinalysis)
This is done on a regular basis to check for infection and/or kidney damage.

X-Rays
X-Rays are used to see if there is an infection in the lungs and to look at bones which may be damaged by sickle cell disease.

Health Care Providers

Audiologist
The person who tests your child's hearing.

Cardiologist (heart specialist)
The doctor who does special tests to check your child's heart.

Child Life Specialist
A person who uses play with your child to lower anxiety and promote understanding and healthy patterns of development during hospital stays. The child life specialist supervises programs for groups of children in the playroom and for the individual child at bedside.

Dentist
The doctor who helps your child keep his teeth healthy and clean.

Gastroenterologist
The doctor who specializes in problems of the liver, stomach, and intestines. This doctor is the person who would perform a liver biopsy.

Genetic Counselor
A person who explains how your child inherited sickle cell disease and your chances of passing it on to future children. They also can tell you if there are any other hereditary disorders that might be in your family. The genetic counselor teaches you and your child about the disease and how to deal with it.

Hematologist (blood specialist)
The doctor who treats your child's sickle cell disease.

Nephrologist (kidney specialist)
The doctor who treats your child's kidneys if they are damaged.

Neurologist
This is a doctor who is an expert in the nervous system. The nervous system includes the brain. This specialist checks for seizures, strokes or related problems.

Neuropsychologist

This is a psychologist who helps evaluate and treat children with learning problems.

Nurse Practitioner
A nurse who has special training that may see your child instead of a doctor at routine medical visits. A nurse practitioner can coordinate care between your pediatrician and hematologist and make sure that things go smoothly if your child is in the hospital.

Nutritionist
The person who gives you advice on the best foods for your child and helps you find ways to get your child to eat what is good for him.

Occupational Therapist
The person who takes your child to "O.T." (Occupational Therapy) when he is in the hospital. These therapists help your child do activities that are useful as well as interesting. These activities can include dressing, cooking, sewing, computer games, etc. These activities can help your child focus on other things besides being sick or in pain.

Ophthalmologist (eye specialist)
The doctor who checks your child's eyes for any sickle cell damage. This doctor can use surgery or laser therapy to correct problems in and around the eye.

Orthopedist (bone specialist)
The doctor who treats damaged bones or joints.

Pediatrician
A doctor who provides medical care for children. A pediatrician can give your child routine care, referring your child to other specialists as needed.

Physiatrist
A doctor who has specialized in the function of the bones, muscles, and nervous system. A physiatrist can help people with special exercises to relieve pain and increase activity.

Physical Therapist
The person who takes your child to "P.T." (Physical Therapy) when he is in the hospital. These therapists also bring hot packs to the clinic if your child is being treated for pain or provide activities designed to help relieve pain, such as mild exercise or whirlpool (hot tub) treatments.

Primary Care Physician
A doctor who provides medical care for people of all ages, from babies to older adults. A primary care physician can give your child routine health care, referring to other specialists as needed.

Psychiatrist
A doctor who has special training to help people deal with stress and can prescribe medications to help relieve stress.

Psychologist
A person who provides counseling for families and children. Psychologists are trained to help children and families deal with a chronic illness, including stress and pain. They also help with other concerns, such as school problems, behavior problems, and depression.

Social Worker
The person who helps families cope better with sickle cell disease. A social worker can help with billing and health insurance, getting medical care in and out of the hospital, and finding support for your emotional needs. You and your child can also talk to a social worker about any of your problems or

TRAVEL LETTER

Re: _____
MR# _____ SAMPLE
DOB: _____

To Whom It May Concern:

_____

is a _____ year old with hemoglobin _____ disease who is followed at

_____ under the care of _____.

In order to decrease the morbidity from their disease, our patients and their families are educated to recognize the symptoms and seek immediate treatment of the following emergencies seen commonly in sickle cell disease. We would appreciate your cooperation in the event that any of these patients come to you for treatment.

1. **Fever greater than 101º F**: Aggressive evaluation for the source of such a fever in the child with sickle cell disease is very important. This evaluation should include CBC, reticulocyte count, blood culture, chest x-ray and urine culture. He or she should be started on IV parenteral antibiotics pending blood culture results.

2. **Acute chest pain or difficulty breathing**: The patient should have a chest x-ray, CBC, reticulocyte count, and consider blood gas studies if there is any evidence of acute respiratory distress. If the patient is febrile, antibiotics should be started. In a patient with severe chest pain and/or a positive chest x-ray, hospitalization is mandatory.

3. **Acute pain not relieved by acetaminophen, fluids, bedrest**: An aggressive evaluation for the source of the pain is mandatory. CBC, reticulocyte count, and other appropriate laboratory tests are also recommended. Follow-up in twenty-four hours to evaluate status.

4. **Marked lethargy or tiredness**: Physical examination documenting the size of the spleen, CBC, reticulocyte count and observation are recommended to rule out splenic sequestration and acute aplastic episode.

5. **Vomiting, diarrhea leading to dehydration**: If the patient cannot take oral fluids, the patient should be hydrated.

6. **Neurologic symptoms (seizures, weakness in the arms or legs, severe headaches, marked dizziness or visual changes)**: The patient should undergo an extensive neurological examination. All patients with neurological symptoms should be admitted to the hospital. The possibility of a cerebrovascular accident should always be considered. Febrile patients demand a spinal tap. Exchange transfusion should be considered, and we should be notified of such a situation immediately.

Telephone number _____ .

Please call Dr. _____ available at telephone number _____ during regular business hours to provide further information about individual patients, to answer any questions, and to screen calls for appropriate physicians. During other times, call the on-call hematologist at

_____ hospital. The switchboard can page one of our physicians

24 hours a day at _____ .

When to Call the Doctor or Nurse

Call to have your child seen right away if your child has one of these danger signs:	**FEVER**	101^0 F or higher
	HEAD/NECK	Severe headache or dizziness Stiff neck
	CHEST	Pain or trouble breathing
	STOMACH	Severe pain and swelling
	COLOR	Loss of normal skin color, very pale or gray
	PENIS	Painful erection
	BEHAVIOR	Seizures Weakness or paralysis (can't move arm or leg) Can't wake up Limping without pain

If you can't reach your doctor, take your child to the Emergency Room.

Call for advice if your child has one of these problems.	**STOMACH**	Vomits more than once Has diarrhea more than once
	FEVER	100^0 F which lasts more than 24 hours
	COLOR	Jaundice (eyes or skin look yellow)
	ARMS, LEGS AND BACK	Pain with no other symptoms
	CHEST	Coughs without fever or chest pain
	NOSE	Runny or stuffy nose
	BEHAVIOR	Acts strangely Refuses to take medicine Refuse to eat or drink Less active than usual

Temperature Conversion Chart

Celsius (°C) to Fahrenheit (°F)

	°C	°F
	36.0	96.8
	36.2	97.2
Normal armpit temperature	**36.4**	**97.5**
	36.6	97.9
	36.8	98.2
Normal temperature by mouth	**37.0**	**98.6**
	37.2	99.0
	37.4	99.3
Normal temperature by rectum	**37.6**	**99.7**
	37.8	100.0
	38.0	100.4
	38.2	100.8
Call your doctor if your child has a fever over 101°	**38.4**	**101.1**
	38.6	101.5
	38.8	101.8
	39.0	102.2
	39.2	102.6
	39.4	102.9
	39.6	103.3
	39.8	103.6
	40.0	104.0
	40.2	104.4
	40.4	104.8
	40.6	105.1
	40.8	105.4

Letter to Schools About Physical Education

Date:

RE: Patient _____

MR# _____

DOB: _____

To Whom It May Concern:

The above-named patient is a _____-year-old followed at _____
(Clinic or Hospital)
for sickle cell disease. She/he is capable of normal participation in a general physical education program, and we would encourage this as much as possible.

However, because of his or her sickle cell disease, there are times when the patient may experience pain in the limbs, shortness of breath or other symptoms with strenuous physical activity. She/he should be excused from such activity at these times, but alternatives should be sought which will allow as much participation as possible in group activities, without forcing the child beyond these temporary limitations.

If you have any questions, please feel to contact me at _____
(Clinic or Hospital)

Sincerely,

_____
(Doctor or Nurse)

_____
(Telephone Number)

California Parent and Adult Support Groups

Sickle Cell Disease Foundation of California
6133 Bristol Parkway, Suite 240
Culver City, CA 90230-6635
Phone: 310-693-0247
Toll Free: 877-288-2873
Fax: 310-693-0266
Website: www.scdfc.org

Sickle Cell Organization of the Inland Counties
2060 University Avenue, Suite 206
Riverside, CA 92507-5210
Phone: 909-684-0420
Fax: 909-684-0340

Sickle Cell Community Health Network of Northern California
610 16th Street, Suite 400
Oakland, CA 94612-1283
Phone: 510-628-0610
Fax: 510-628-0611
Website: www.sicklecell.net
E-mail: sicklecellnet@sbcglobal.net

Sickle Cell Disease Association
636 Broadway, Suite 314
San Diego, CA 92101-5410
Phone: 619-263-8300
Fax: 619-233-3557

Children's Hospital San Diego
3020 Childrens Way, MC-5081
San Diego, CA 92123-4223
Phone: 858-966-6709
Fax: 858-966-8991

Miller Children's Hospital Long Beach Memorial Medical Center
Sickle Cell Disease Center
2801 Atlantic Avenue
Long Beach, CA 90801-1737
Phone: 562-933-8610
Fax: 562-933-8610

Sickle Cell Adult Support Groups

Kaiser Inglewood
110 North La Brea Avenue
Inglewood, CA 90301-1766
Phone: 310-419-3451
Fax: 310-419-3370

St. Agnes Hospital
1111 East Spruce Avenue
Fresno CA 93720-3330
Phone: 559-450-5372
Fax: 559-450-3032

Sickle Cell Community Health Network of Northern California
610 16th Street, Suite 400
Oakland, CA 94612-1283
Phone: 510-628-0610
Fax: 510-628-0611
Website: www.sicklecell.net
E-mail: sicklecellnet@sbcglobal.net

Sample Pain Management Agreement

Agreement between the Teen and the Sickle Cell Care Team

GOALS:

1. The teen will perform self-care activities as specified.
2. The teen will remain as pain free as possible.

The Teen's Responsibilities	Staff Responsibilities
1. Will discuss how well the pain medication is working with the health care team.	1a. Will administer the agreed upon pain medication and discuss the treatment plan with the teen. 1b. Will adjust pain medication dosage as needed within safe guidelines.
2. Will drink the needed amounts of fluids.	2. Will provide fluids of choice and encourage intake when necessary.
3. Will do deep breathing exercises every 4 hours or use incentive spirometer as agreed upon.	3. Will encourage and assist as needed.
4. Will get out of bed for 10 minutes during both day and evening shifts and increase this amount of time over time.	4. Will help, when needed, with walking during both day and evening shifts, and provide distraction activities.
5. Will do relaxation exercises for 30 minutes on each shift.	5. Will encourage and assist as needed.

California Children Services (CCS) Approved Sickle Cell Disease Centers

Northern California

U.C. Davis Medical Center
2516 Stockton Boulevard, TICON II
Sacramento, CA 95817
(916) 734-2782
Jonathan Ducore, MD, Director
and Theodore Zwerdling, MD

Children's Hospital and Research Center at Oakland
747 52nd Street
Oakland, CA 94609
(510) 428-3372
Elliot Vichinksy, MD, Director
*Services also provided in
Sacramento, CA

Kaiser Permanente Medical Center
280 W. MacArthur Blvd.
Oakland, CA 94611
(510) 752-1000
Stacy Month, MD, MPH

U.C. San Francisco Medical Center
505 Parnassus Avenue
San Francisco, CA 94143-0110
(415) 476-3831
William C. Mentzer, MD, Director
& Marion Koerper, MD, Co-Director

Packard Children's Hospital at Stanford
725 Welch Road
Palo Alto, CA 94304
(650) 858-1335
Bertil E. Glader, PhD, MD, Director

Children's Hospital of Central California
9300 Valley Children's Place
Mail Stop OS14
Madera, CA 93638
(559) 353-5460
Robert Mignacca, MD, Director

Southern California

City of Hope Medical Center
1500 East Duarte Road
Duarte, CA 91010
(626) 301-8426
Nadia Ewing, MD, Director

Cedars-Sinai Medical Center
8700 Beverly Boulevard, PM 1165
Los Angeles, CA 90048
(310) 423-4423
Carole Hurvitz, MD, Director

Childrens Hospital of Los Angeles
4650 Sunset Boulevard, MS61
Los Angeles, CA 90054
(323) 669-4100
Thomas Coates, MD, Director

Kaiser Permanente Medical Center, So. California Regional Hemoglobinopathy Center
6041 Cadillac Drive
Los Angeles, CA 90034
(800) 734-5155
(323) 857-4462
Elaine E. Smith, MD, Director

LAC/USC Medical Center
1240 North Mission Road, L-902
Los Angeles, CA 90033
(323) 226-3853
Robert Baehner, MD, Director

UCLA Medical Center
Pediatrics, MDCC A2-312
10833 Le Conte Avenue
Los Angeles, CA 90095-1752
(310) 825-6447
Stephen A. Feig, MD, Director

Harbor/UCLA Medical Center
1100 West Carson Street, Box 468
Torrance, CA 90509
(310) 222-4154
Lance Sieger, MD, Director

Miller Children's at Long Beach Memorial Medical Center
2801 Atlantic Avenue
Long Beach, CA 90801
(562) 492-1062
Paula Groncy, MD, Director

Loma Linda University Medical Center
11234 Anderson Street
Loma Linda, CA 92354
(909) 558-2283
Antranik Bedros, MD, Director

Children's Hospital of Orange County
455 S. Main Street
Orange, CA 92668
(714) 532-8459
Diane Nugent, MD, Director

U.C. Irvine Medical Center
101 City Drive, Building 27
Orange, CA 92668
(714) 456-6615
Stanley Caleerwood, MD, Director

Children's Hospital and Health Center of San Diego
3020 Children's Way
San Diego, CA 92123-4282
(858) 966-5811
Faith H. Kung, MD, Director

Chances of Having a Baby with Sickle Cell Disease

Both Parents Have Sickle Cell **Disease**

Sickle Cell Disease
(SS)

Sickle Cell Disease
(SS)

Sickle Cell Disease (SS) | Sickle Cell Disease (SS) | Sickle Cell Disease (SS) | Sickle Cell Disease (SS)

When both parents have sickle cell disease, **all** their children will also have the disease.

One Parent Has Sickle Cell **Disease** and **One** Parent Has Sickle Cell **Trait**.

Sickle Cell Disease
(SS)

Sickle Cell Trait
(AS)

Sickle Cell Trait (AS) | Sickle Cell Trait (AS) | Sickle Cell Disease (SS) | Sickle Cell Disease (SS)

When one parent has sickle cell disease and the other has a trait, they have a 50% chance (or 1 out of 2) of having a baby with sickle cell disease. Each time they get pregnant, they have the same chance.

One Parent has Sickle Cell **Disease** and **One** Parent Has **No Hemoglobin Trait**

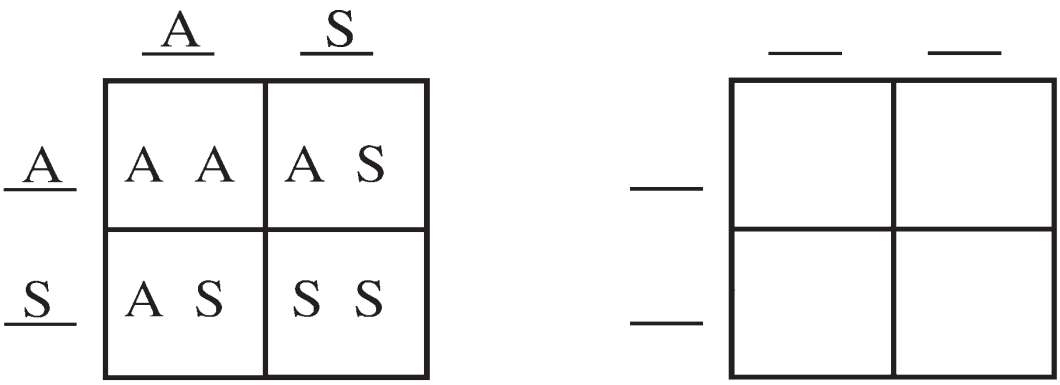
No Trait
(AA)

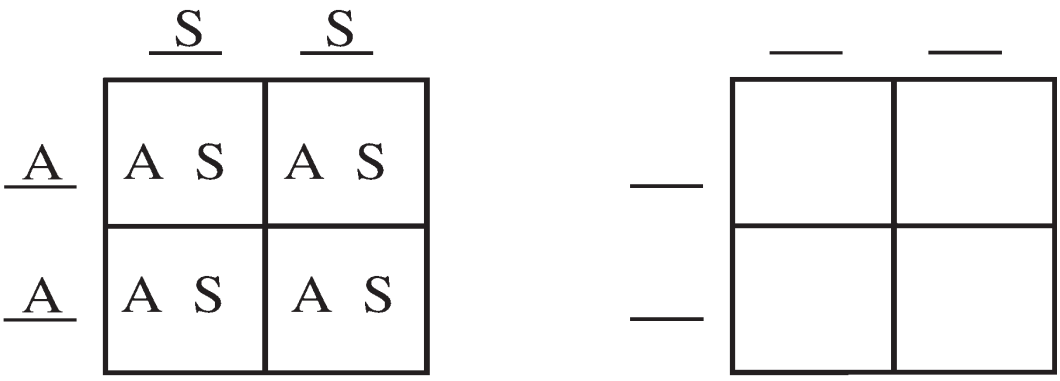
Sickle Cell Disease
(SS)

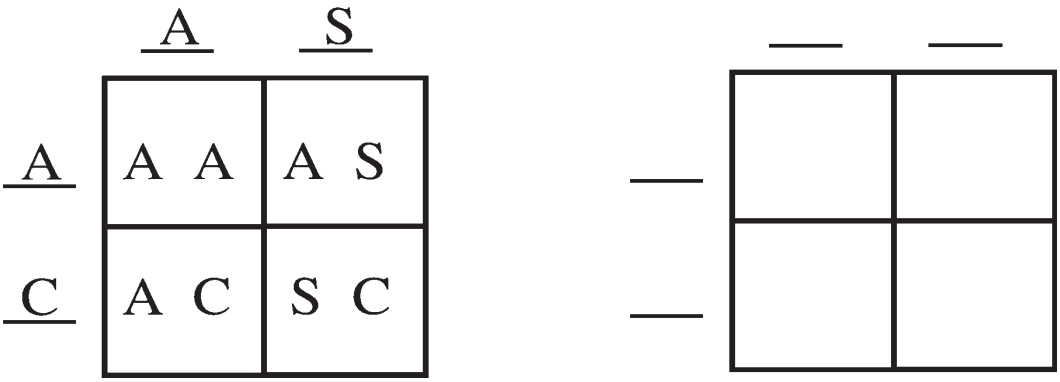
Sickle Cell Trait (AS) | Sickle Cell Trait (AS) | Sickle Cell Trait (AS) | Sickle Cell Trait (AS)

When one parent has sickle cell disease and the other has **no** trait, the couple **cannot** have a baby with sickle cell disease. Each of their babies will have a trait.

Diagrams of Inheritance *(Punnett Squares)*







What is a 504-Plan?

A 504-plan is a legal, binding document that began with Section 504 of the Rehabilitation Act of 1973. It is designed to help any student attending public school who has special physical (such as seizures, diabetes or sickle cell disease) or psychological (such as attention deficit disorder) needs to feel comfortable in the regular classroom.

Who is eligible?
Your child is eligible if he is considered disabled. This is a term that includes any condition that might cause your child to be unable to perform "major life activities" with the rest of the class. Where sickle cell disease is concerned, these activities include having to wait with the rest of the class to go to the bathroom or get a drink of water; having to have assignments or tests done quickly, even though your son might feel fatigued because of sickle cell disease; having to go out for recess in extreme temperatures; and having to run without stopping in PE, even though your child has trouble breathing or is tired because of sickle cell disease.

How do I get a 504-Plan?
The first step is to contact the school principal and fill out a referral form. Requests have to be in writing, not just verbal. You will need a letter from your physician describing exactly the kinds of adjustments that are needed because of sickle cell disease and why. Then, a meeting is scheduled between you and your child's teachers, counselors, principal, support staff as needed (e.g., school nurse) and your child. After discussion, a 504-Plan is developed that will list the special accommodations. When everyone agrees with the list, the plan is signed and sent to all of your child's teachers, who are legally required to comply.

Does a 504-Plan become part of my child's permanent record?
No, some 504-Plans are temporary. Your child may not always need special accommodations because the concerns that come with sickle cell disease can change from school year to school year. Not all schools will provide a temporary 504-Plan because it is so legally binding. Check with your school for local policy.

What kinds of accommodations are available?
Depending on the type and severity of your child's sickle cell disease related problems, the school may consider any of the following options and can even make up new accommodations to fit your child's needs. In addition to those already mentioned, the 504-Plan may recommend:
- Extended time for testing
- Extra time for moving between classes
- Use of a journal for communication between student/counselor/parents
- Special transportation to school and field trips
- Allowance for health-related absences
- Time in the school nurse's office so that pain can be managed at school and your son does not have to go home every time he has pain

Does a 504-Plan cost anything?
Not directly. Your taxes pay for the special transportation or other building changes such as ramps. Some federal grant money is also available for meeting the needs of 504-Plan students. As parents, you pay nothing to have your child's 504-Plan needs met.

How can I get more information?
As part of the Rehabilitation Act of 1973, this section can be seen at this website: www.dese.state.mo.us/divspeced/staccess.html. Also, most schools have a copy of the 504-Plan regulations in their counseling office.

Resources — *Books, Foundations, Information and Support*

National Foundations

Sickle Cell Disease Association of America, Inc.
www.sicklecelldisease.org

The American Sickle Cell Anemia Association
www.ascaa.org

National Heart, Lung, and Blood Institute
(301) 592-8573
www.nhlbi.nih.gov/index.htm

Annie E. Casey Foundation
www.aecf.org

March of Dimes
www.marchofdimes.com

Brave Kids - Help for Children With Chronic, Life-threatening Illnesses or Disabilities
www.bravekids.org

Starbright Foundation
www.starbright.org

Medical Home (American Academy of Pediatrics) Particular focus on transition to adult care.
www.medicalhomeinfo.org

Resources for Information

GeneHELP
State of California
Department of Health Services
Genetic Disease Branch
850 Marina Bay Parkway, F175
Richmond, CA 94804
(510) 412-1502

MEDLINEplus
www.nlm.nih.gov/medlineplus/sicklecellanemia.html

Sickle Cell Anemia Mini-Course and Virtual Lab
http://k14education.uams.edu/scvlab

Support Organizations

Genetic Alliance
www.geneticalliance.org

National Organization for Rare Disorders
www.rarediseases.org

Genetic and Rare Diseases Information Center
P.O. Box 8126
Gaithersburg, MD 20898-8126
Phone: (888) 205-2311
TTY: (888) 205-3223
E-mail: gardinfo@nih.gov
Fax (240) 632-9164

The Family Village
www.familyvillage.wisc.edu

California Department of Rehabilitation
www.dor.ca.gov

For more information on parenting:

Beal, Anne C., Villarosa, Linda, and Abner, Allison, *The Black Parenting Book: Caring for Our Children in the First Five Years*, Broadway Books, 1998.

Poussaint, Alvin S., and Comer, James, *Raising Black Children*, Plume, 1992.

Price, Hugh, *Achievement Matters: Getting Your Children the Best Education Possible*, Kensington Publishing Corporation, 2003.

Boyd-Franklin, Nancy and Franklin, A. J., PhD, *Boys Into Men: Raising Our African American Teenage Sons*, Penguin Group, 2001.

Wright, Marguerite, *I'm Chocolate, You're Vanilla: Raising Healthy Black and Biracial Children in a Race Conscious World*, Jossey-Bass, 2000.

Ginott, Haim, and Ginott, Alice, Goddard, H. Wallace, *Between Parent and Child*, Three Rivers Press, 2003.

Corwin, Donna G., *The Tween Years: A Parent's Guide for Surviving Those Terrific, Turbulent and Trying Times*, McGraw-Hill, 1998.

Rodriguez, Gloria, *Raising Nuestros Niños: Bringing Up Latino Children in a Bicultural World*, Simon & Schuster Adult Publishing Group, 1999.

Tanen-Leff, Patricia and Walizer, Elaine H., *Building the Healing Partnership: Parents, Professionals, and Children with Chronic Illnesses and Disabilities*, Brookline Books, 1993.

Glossary

Acute Chest Syndrome

Pneumonia caused by infection and/or sickle cell damage in the lungs. Signs of acute chest syndrome may include fever, chest pain, coughing, shortness of breath, or difficulty breathing. This is a medical emergency.

Anemia (low blood)

A condition in which there is less hemoglobin in the blood than usual so that the blood can't carry as much oxygen.

Aplastic Episode

An episode when the bone marrow stops making red blood cells. The blood count may fall much lower than usual. If it happens, it is usually with a fever or infection.

Avascular Necrosis

This is the term for damage to joints caused by sickle cell disease.

Carrier

A person who has one gene for Hemoglobin A and one gene for another type of hemoglobin. This person is also referred to as having a hemoglobin trait. A carrier doesn't have the disease, but two carriers can have a baby with sickle cell disease.

Chromosome

Structures containing the genes in the body. Most people have 46 chromosomes. Prenatal testing can be done to study an unborn baby's chromosomes.

Complete Blood Count (CBC)

A blood test which measures the size of the red blood cells and the amount of hemoglobin. It tells the number of red blood cells, white blood cells, and platelets.

Cross-Matching Blood

Before a blood transfusion, the blood bank checks to see if the donor blood is a good match with the blood of the person who will receive it. Many different proteins on the blood cells can be checked by "phenotyping" in order to find the best match and prevent complications from the transfusion. A cross-match is done with each transfusion.

Dehydration

A condition caused by not having enough water in the body. Dehydration can happen with diarrhea, fever or exercise. It may cause a sickling episode in someone with sickle cell disease.

Electrophoresis

One of the best blood tests to find out a person's hemoglobin type. It shows most hemoglobin traits and can determine different types of sickle cell disease.

Exchange Transfusion

Blood transfusions can be given through an intravenous line (IV) to increase the hemoglobin to a normal level, or can be done with a machine, which removes the sickle cell blood and replaces it with blood from donors. An exchange transfusion requires two IV lines: one to take the sickle blood out and one to transfuse the donor blood in.

Gene

The basic unit of heredity. Genes are passed on by a mother in the egg and by a father in the sperm. People have about 35,000 genes which determine many characteristics, including hemoglobin type.

Hemoglobin

The substance which carries oxygen in red blood cells. People with sickle cell disease often have lower hemoglobin levels.

Infarct

A blockage of blood flow that causes tissue to die because it doesn't have enough oxygen.

Inherited

A characteristic passed on from parents to their children. Sickle cell disease is an inherited disease.

Jaundice

Yellowish color of the skin or eyes. It is caused by coloring materials from red blood cell breakdown.

Leg Ulcer

A breakage in the skin that begins as a small sore on the lower leg (above, over and/or around the ankle). It can be caused by injury and decreased blood flow.

Neuropsychological Testing

This is testing done in young children or adolescents to determine how to help them if they are having a hard time in school. A specially trained psychologist will ask the child questions; there may be some questions for the child to read and then write answers. Sometimes testing can take as long as eight hours and has to be done in two sessions.

Glossary (cont.)

Pain Medication Dependency

Long-term use of some medication scan cause the body to become used to the medication, so that stopping it suddenly causes discomfort. When someone has taken pain medication for a long time and the pain has stopped, the doctor will gradually decrease the dose of medication in order to prevent such problems. Dependency is not the same as addiction.

Pain Medication Tolerance

Some medications become less effective if they have to be used for a long time. When that happens, higher doses are given to decrease the pain.

Priapism

A persistent, painful, unwanted erection of the penis caused by sickling.

Prophylactic Penicillin

Penicillin which is given in order to reduce the number and severity of infections in children with sickle cell disease.

Pulmonary Hypertension

This is a term for the heart having to pump harder than usual to get blood to the lungs. In sickle cell disease it is caused by lung damage from sickling. An echocardiogram will help determine if pulmonary hypertension is present.

Retinopathy

Damage to the back of the eye (retina) caused by blockage in the small blood vessels and scarring in that area. This can lead to poor vision and even blindness. After the age of ten years every sickle cell patient should see an eye doctor once a year.

Sickle Cell Anemia

Another name for SS disease, the most common type of sickle cell disease.

Sickle Cell Disease

A term which refers to all types of sickle hemoglobin disorders, such as SS disease, SC disease and S beta thal disease.

Splenectomy

Surgery to remove the spleen. This is done to cure the serious anemia that happens when blood is trapped in the spleen.

Spleen

An organ on the left side of the body that may be felt below the rib cage. It is a filter to remove bacteria from the blood. This organ does not work well in sickle cell disease. It can trap blood and become enlarged.

Splenic Sequestration

One type of episode that can occur in patients with sickle cell disease which can be life threatening. It is caused by blood being trapped in the spleen.

Trait (see Carrier)

Transfusion

Blood given to someone because of a very low blood count, to prepare for surgery or to treat certain complications of sickle cell disease.

Vaso-Occlusive Episode

Occurs when sickle cells block the flow of blood. This causes pain and, if severe, tissue damage.